

BIOMICROSCOPY OF THE EYE

Slit Lamp Microscopy of the Living Eye

VOLUME II



FRONTISPIECE. Biomicroscopic view (direct focal illumination) with contact lens, through cornea, lens, vitreous, and fundus.

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Slit Lamp Microscopy of the Living Eye

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VOLUME II



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BIOMICROSCOPY OF THE EYE

VOLUME II

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E-Y

PUBLISHED IN 1949

To
MY WIFE

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BIOMICROSCOPY OF THE EYE

Slit Lamp Microscopy of the Living Eye

Chapter Eighteen

THE IRIS

THE importance of careful biomicroscopy of the iris cannot be overemphasized. The iris is composed of both ectodermal and mesodermal derivatives, and all the defects, both developmental and acquired, that affect such tissues are observed. From the standpoint of genetics, iris studies have proved extremely fruitful. The classical examples of hereditary iris color variations, so ably analyzed by Davenport according to Mendel's law, are discussed later. Vogt, in his observations on uniovular (identical) twins, has brought out conclusive evidence that certain changes, e.g., senile disturbances of the pupillary pigment border and stroma, can be explained only on a genetic basis. Biomicroscopy of the iris can disclose early many intrinsic changes attributable to abiotrophy and senility. Because of the frequency of iridic involvement in systemic disease, its value in the study of such pathologic complications is inestimable. From the standpoint of function there is no better method than biomicroscopy for the study of pupillary mobility. Frequently a pupil that appears rigid by ordinary examination will disclose a local or sectorlike reaction to light. Together with gonioscopy this method affords the best means of observing the role that the iris plays in the interference with the intra-ocular pressure mechanism.

The anterior iris face, with its markings exposed like the dial of a clock, lends itself ideally to biomicroscopic inspection, except in the extreme periphery where it is hidden by the overlapping sclera.* Even before the days of biomicroscopy, the ease with which the surface of the living iris could be inspected, by means of the loupe or binocular microscope, led to very accurate descriptions of its

* Gonioscopy reveals parts of this hidden portion (see Volume I, Chapter 17).

PLATE XLI

FIG. 1. Normal brown iris, illustrating the pigment seam, crypts, contraction furrows. (Direct focal illumination.)

FIG. 2. Normal blue iris, illustrating surface markings. (Direct focal illumination.)

FIG. 3. Normal blue iris. (As viewed by optic section.)

FIG. 4. Normal blue iris, showing physiologic rarefaction below.

FIG. 5. High-power view of Figure 4.

FIG. 6. Blood vessels seen in normal iris.

FIG. 7. Blue iris as seen by daylight.

FIG. 8. Same iris as shown in Figure 7 observed biomicroscopically in diffuse illumination. Brownish color is due to reflection from the posterior pigmented surface.

FIG. 9. Tongue of iris stroma adhering to the seam. (Pupillary pigment border.)



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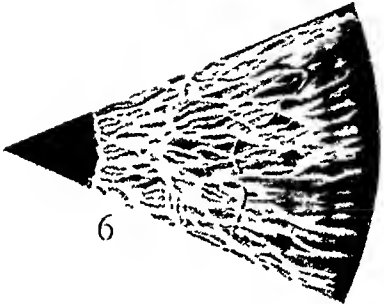
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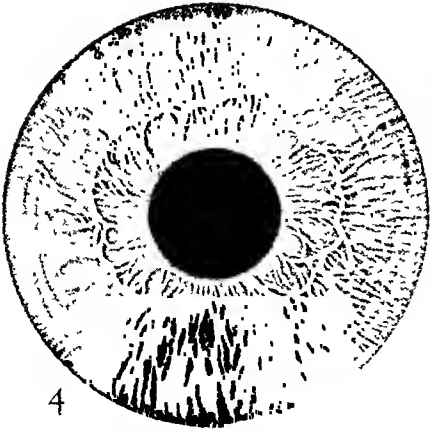
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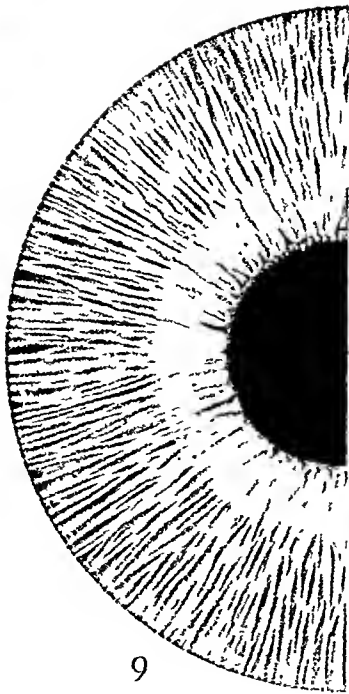
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architecture or surface markings, its luster and color. The structure of the iris, composed of tissue derived both from ectodermal and mesodermal elements, is optically heterogeneous in character.* Normally, the complex structure retards internal illumination or retro-illumination (transillumination) of its substance and thus renders it more obstructive to these types of illumination than do the tissues of the so-called transparent ocular media.

Owing to its heavily pigmented posterior face it is impossible to transilluminate or retro-illuminate even the normal blue or gray iris with light reflected from the lens or fundus. The added surface pigmentation of the normal brown iris further obstructs this type of illumination. Only albinotic or atrophic irides transmit such light readily. However, employing the narrow beam of direct focal illumination in light blue or gray irides, it is possible, within limits, to obtain an optic section through their substance. Also by indirect lateral illumination (a form of scatter) due to internal dispersion, the sphincter or such changes as hemorrhages (suggilation), which usually are not visible in direct focal or diffuse light, may be observed within the tissue of the iris. The normal variations in coloring and architecture are so multitudinous (no two irides are ever alike) that the necessity for observing many cases is evident. Not only are there variations in coloring and architecture in different persons but even in the two eyes of the same person, and a given iris undergoes changes in color and structure with the passage of time from childhood to senility.

Both blue and brown irides may reveal well-developed surface markings (Plates XLI, figs. 1, 2; XLII, figs. 1, 2, 3, 4, 5). But some blue-gray irides have such a compact structure that relatively few markings are visible. Heavily pigmented brown irides may present a uniformly structureless spongelike surface without trabeculae. In some of these cases a sharp line may indicate the declination (lesser circle) of the pupillary zone toward the pupillary margin. Even in the irides of Negroes, the ciliary portion may disclose this type of

* In comparison, the cornea, lens, and vitreous, composed of "gel-like" transparent tissue, can almost be considered optically homogenous. Ordinary clear glass would be an example of a substance having the property of true optical homogeneity.

uniform spongy structure while the pupillary zone of the same iris may show a trabecular design with many crypts. When the surface of a dark brown spongy type of iris is studied under high power, an intricate and delicate pigmented network can just be made out with difficulty. This network may represent starlike branchings of the melanophores. The morphology and biomicroscopic appearances will now be considered in more detail.

MORPHOLOGY*

Histologically, proceeding internally, the iris may be divided into five layers: (1) endothelium; (2) anterior border layer; (3) vessel layer; (4) dilator pupillae, composed of (a) posterior border lamella and (b) pigmented spindle cells; and (5) pigmented epithelium (Fig. 306).

Endothelium. Although difficult to demonstrate in preparations, the endothelium is generally accepted as being present in the form of a poorly defined noncontinuous layer. It is important to note that it does not cover the openings of the crypts and that it is poorly developed in the neighborhood of the lesser circle.

Anterior Border Layer. The anterior border layer is thin, dense and avascular, composed chiefly of melanophores (in the brown iris), and sparsely interspersed collagenous fibrillae. This layer is missing at the entrance of the crypts and is thin over the floor of the contraction furrows. Its density and pigment content determine the color of the iris. Salzmann⁵⁹¹ states: "Blue irides have a delicate border layer and almost unpigmented cells; brown irides have a thick border layer and very heavily pigmented cells." At the periphery, the anterior border layer and the underlying stroma are thrown into folds (contraction furrows). The anterior border layer is thinnest in the troughs of these folds.

Vessel Layer. The vessel layer forms the larger part of the iris stroma and contains numerous larger vessels (most of them coursing in a meridional direction) and nerve plexuses. The stroma itself consists mainly of delicate loose fibrillae branching chiefly in a

* For a more detailed discussion of the anatomy and histology of the iris, the reader is referred to Salzmann's "Anatomy and Histology of the Human Eyeball."⁵⁹¹

meridional direction. In the pupillary zone and toward the periphery of the ciliary zone many of the fibrillae take a circumferential direc-

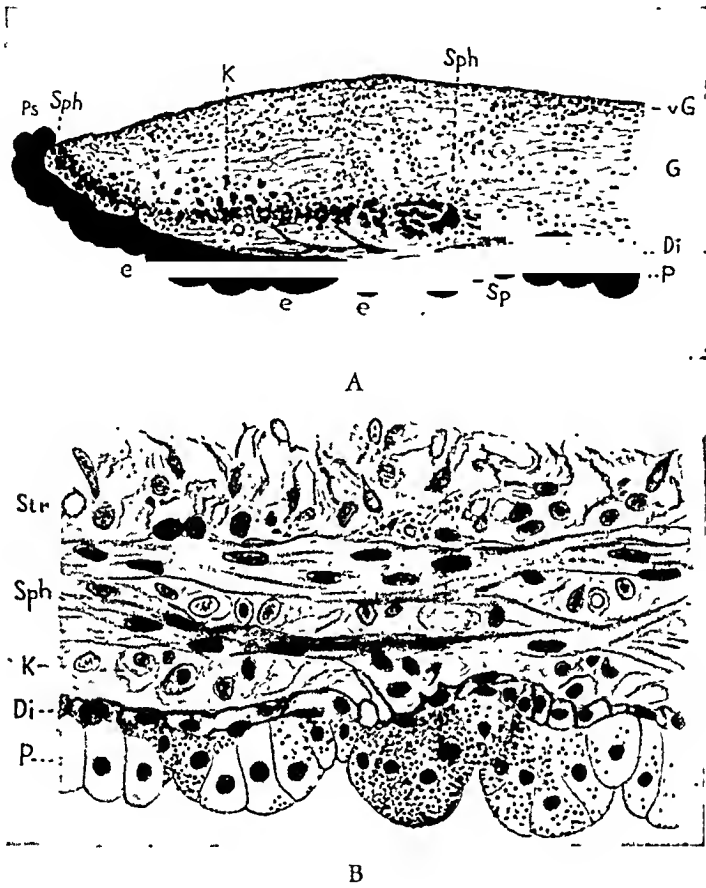


FIG. 306. Histologic section of iris showing its layers. A. Pupillary zone of the iris, meridional section, $\times 60$. *vG*, anterior border layer (with endothelium); *G*, vessel layer; *Di*, dilator pupillae; *P*, pigment epithelium; *Sph*, sphincter pupillae; *e*, radiation of the dilator into the sphincter from behind; *Ps*, pigment seam of the pupillary border; *K*, clump cells. (After Salzmann.) B. Ectodermal layers of the posterior iris surface, transverse section, bleached, $\times 330$. *Str*, stroma of the vessel layer of the iris; *Sph*, sphincter pupillae; *K*, clump cell; *Di*, dilator pupillae; *P*, pigment epithelium. (After Salzmann.)

tion. The stroma also contains melanophores, clump cells (Koganei), and the sphincter, the latter two being derived embryologically from the posterior or ectodermal layers. Both pupillary zone and ciliary zones contain crypts. They appear deeper, larger, and more undermined in the pupillary zone. The sphincter lying in the deep stroma has the form of an annular band about 1 mm. wide. Its border is intimately connected with the pigment of the pupillary border (seam) while its peripheral boundaries and under surface have direct connection with the underlying dilator pupillae.

The stromal and vessel layer is thinnest at the root of the iris and

is thickest in the pupillary zone near the lesser circle. The latter area (near the sphincter) is the most highly vascularized, and it is only here, according to Kruckmann,⁵¹⁶ that a real capillary network exists.

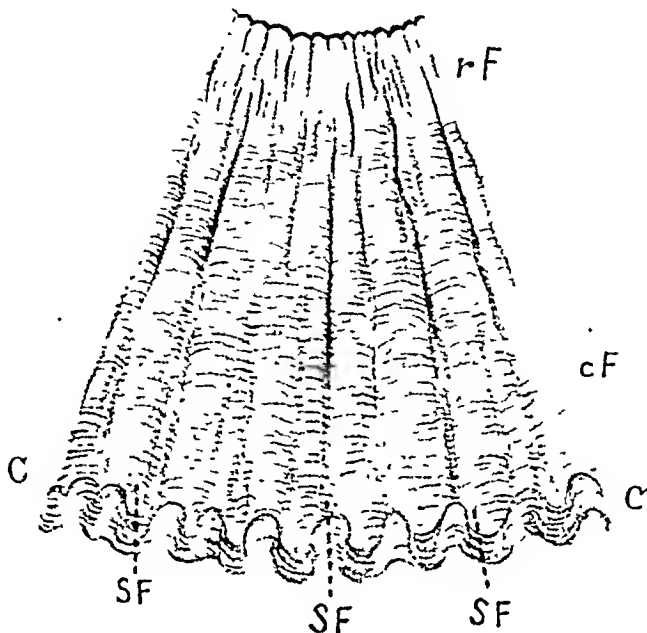


FIG. 307. Relief of a posterior iris surface, under reflected light, X9. *rF*, radial furrows of the pupillary zone; *sF*, structural furrows; *cF*, circular furrows; *C*, anterior border of the corona ciliaris. (After Salzmann.)

In other parts there are some small capillaries but not in the form of a continuous net. This fact is evidenced under certain pathologic conditions, e.g., roseolae in which congestion of the superficial capillaries appears as disconnected stars or rosettes.

Dilator Pupillae. The dilator pupillae is composed of a strongly fused double layer, the deeper being pigmented. It lines the under surface of the vessel layer, extending from the iris root to the edge of the sphincter. Embryologically the dilator muscle is derived from the layer of pigmented epithelium.

Pigmented Epithelium. The pigmented epithelium is the layer of cylindric cells lining the posterior face of the iris. These cells are so densely pigmented that details concerning them can only be seen in depigmented preparations. This layer represents the continuation

of retinal-derived ciliary epithelium over the posterior iris surface. In meridional section one sees a profile of the system of circular furrows characteristic of the markings of the posterior iris surface. These markings can best be seen by inspection of a flat (frontal) anatomic preparation of the entire dark brown or black surface. Both radial and circular markings or folds can be distinguished on this posterior aspect. The latter are much finer and are found only in the region of the structural-furrow system where they cross them in a regular manner. There are two systems of radial folds: One is composed of fine, shallow grooves in the pigmented epithelium, which extend to the pupillary border and continue around it (Fig. 307). This produces the normal beading (ruff) or pupillary excrescences (seen from in front) of the pupillary border. In the second, the structural furrows are deeper at first and then flatten out as they approach the ciliary border. They begin about 0.5 mm. from the end of the pupillary system of folds. This design on the posterior surface of the iris is of interest to the biomicroscopist because its existence may explain certain alterations found on the lens capsule, e.g., detachment of superficial lamellae of the lens capsule (page 1223).

METHODS OF EXAMINATION

Diffuse Illumination. The very nature of the iris structure enhances its examination in diffuse illumination. The whole surface may be viewed in the light of the pre- or postfocal (unfocused) beam. In order to obtain a field large enough to include the entire iris surface, it is necessary to use low magnification. In this way it is possible to make a survey of the entire iris surface (Plate XLII, figs. 1-8). At a glance not only the normal or any gross structural or color changes of the stroma but also the iris mobility may be seen. This form of illumination is recommended for use at the beginning of all biomicroscopic examinations of the iris.*

* It may be advisable to employ a miotic in certain cases where extreme "stretching-out" of the iris is desirable. This may be of value in sensitive subjects by diminishing the amount of light which enters the pupil.

PLATE XLII

FIG. 1. Dark brown iris. (Diffuse illumination.) Observe paucity of surface markings and note the lighter color in the bottom of the crypts and contraction furrows.

FIG. 2. Blue iris, illustrating the delicate trabecular structure.

FIG. 3. Grey iris.

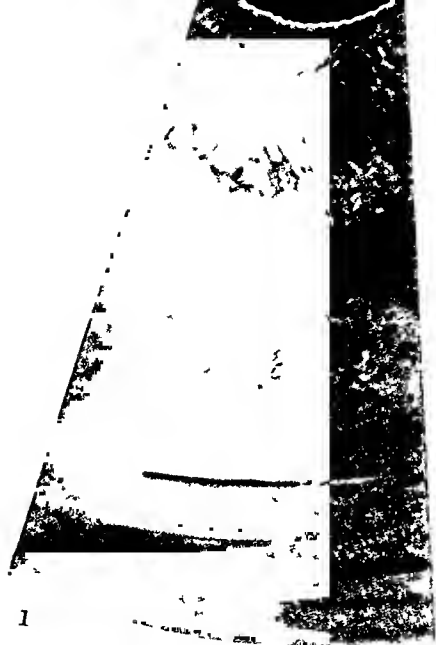
FIG. 4. Blue iris. Large crypts. Well-formed lesser circle.

FIG. 5. Brown iris with well-formed lesser circle. Wolffian bodies.

FIG. 6. Iris bicolor.

FIG. 7. Piebald iris.

FIG. 8. Iris bicolor. Superficial leaf is brownish. Deeper leaf, greenish blue.



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*Direct Focal Illumination.** When the focal point of the beam is directed obliquely toward the iris, an area of highly illuminated surface is seen, corresponding in shape and size to that of the slit opening-

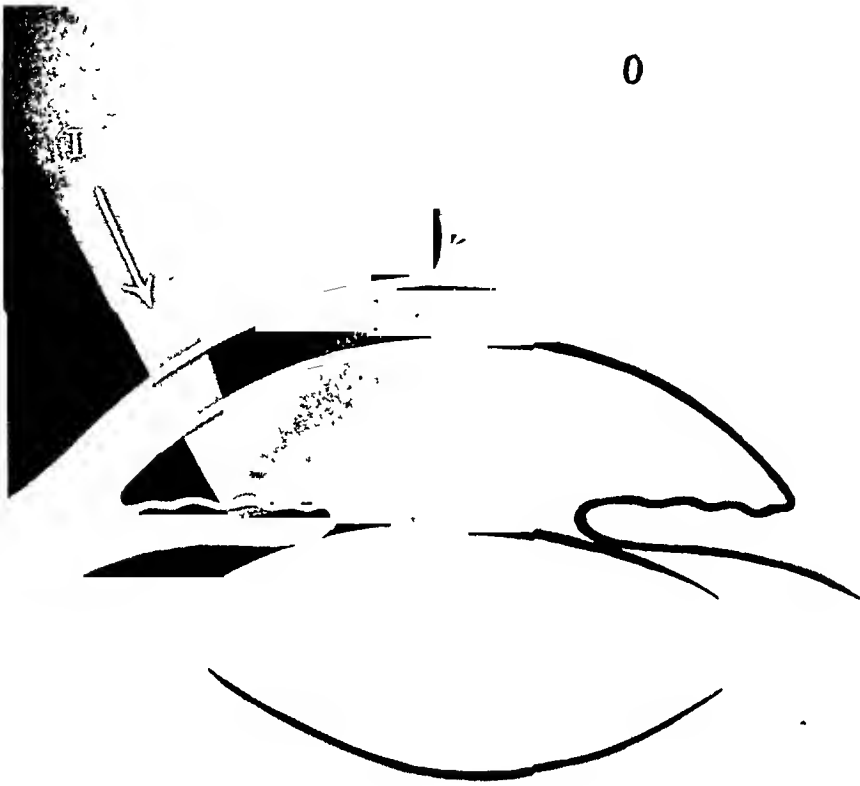


FIG. 308. Diagram illustrating the technique of direct focal illumination of the iris. *I*, axis of illumination; *O*, axis of observation.

ing (Fig. 308). With the widest opening of the slit, the highest intensity of illumination will result (Plate XLI, figs. 1, 2). It is generally advisable to have the focused light beam directed from the temporal side and to employ a wide angle (from 45 degrees to 90 degrees) between the axes of illumination and observation. (Because of the localized area of illumination higher magnifications for observation can be employed.) However, contrary to what occurs in the

* Koepe has recommended the use of a yellow filter which helps to cut down on dazzling and tends to bring out fine details in greater relief. He found also that the blue or red-free filter assists in differentiating between light brown pigment (melanophoric?) and dark brown pigment (retinal). With this filter the yellow color of the light brown pigment is marked so that it becomes indistinguishable from the surrounding background, while dark brown pigment appears black.

cornea and lens, no sharp block (parallelepiped or wedge) will form. Such an optical phenomenon can only occur (as mentioned before) in tissue that transmits light more readily.

Depending on the texture of the iris, some of the light enters the stroma and permits the visualization of certain details. This is particularly true of blue and gray and of certain brown irides (where the trabecular structure is visible). In many dark brown irides the surface is so densely pigmented that it appears smooth and spongy. In these cases it may be impossible, except for the pigment excrescences at the pupillary border and the contraction furrows, to discern surface markings or any of the trabeculae. Here, a thickening in the region of the lesser circle will be present with a sudden posterior or inward declination of the tissue axially toward the pupillary margin, indicating the pupillary zone. A more gradual sloping of the ciliary zone to the periphery may also be perceived. When crypts are present even in darkest brown irides, the absence of the anterior border layers over their openings may allow inspection of the deeper leaf which, despite the underlying retinal pigment layer, may appear lighter brown or yellow in color.

An inspection of the entire iris should be made. Until the patient has become accustomed to the brilliancy of light of the focal beam, it is advisable first to inspect the peripheral regions and then the pupillary zone because, as the beam approaches the pupillary pigment border, some of the light of necessity passes into the pupil and causes dazzling. Hence, as previously mentioned, the advisability of employing a wide angle between observation and illumination, since with a wide angle more light is directed to the less sensitive parts of the retina. As the pupillary zone is approached not only the region of the lesser circle (frill or collarette) but also the pupillary pigmented excrescences and intervening tissue may be studied to a degree impossible by any other methods. The extreme mobility of the pupillary zone under the influence of the light beam is a striking feature. In lightly colored irides, movements of individual fibers of the deeper layer may be seen following the actions of the sphincter.

In pathologic states, as will be seen later, the method of direct

focal illumination will disclose a host of structural tissue variations as well as alterations due to exudative, vascular, atrophic, and neoplastic changes.

Optic Section. With the narrow beam it is possible, in many instances and within certain limits, to obtain an optic section of the iris tissue (Plate XLI, fig. 3). The exceptions, of course, depend on the density and pigmentation of the stromal tissue. The blue or lightly pigmented iris lends itself best to this form of illumination. Where the narrow beam first contacts the iris surface, a delicate line appears (impact line). This probably represents the surface of the anterior border layer. Immediately below it are the fibers of the trabeculae. At times, when the trabeculae are of loose construction, single or branched fluffy trabecular fibers seem to arch up and contact this impact line. The trabeculae are always enmeshed and connected to one another by diaphanous, almost invisible ground tissue. Occasionally in light colored irides a vessel-bearing trabecula may be seen. In this case the center of the fiber consists of a delicate reddish line. In some cases large or small rounded or elliptical spaces are formed between the fibers. These spaces are situated below the surface and differ from crypts in that they do not have surface openings, since even the most superficially placed ones are always covered by the surface impact line. On the other hand, no tissue is visible over the openings of a real crypt, as indicated by the absence of an impact line. Koeppe, using a yellow filter with indirect illumination, has described a delicate subsurface system of furrows or cavities corresponding to the infoldings of the surface. He stated that they are seen in light irides, especially in older individuals, and that they have a "frosted" appearance. He interpreted them as being lymph-containing spaces which he claimed resulted in the aged from regeneration into vacuoles and cavities of a subendothelial system of vesicles. I have been unable to identify these structures.

It is impossible to tell where the superficial leaf or layer ends and where the deeper leaf starts. One has the impression, however, that the structure of the deeper leaf is more compact. Just under the deeper leaf is the dark yellowish or brown reflection of the posterior

retinal pigment layer. Even when a deep large crypt extends through the entire thickness of the stroma or when there is marked stromal atrophy, it is impossible by optic section to identify the individual

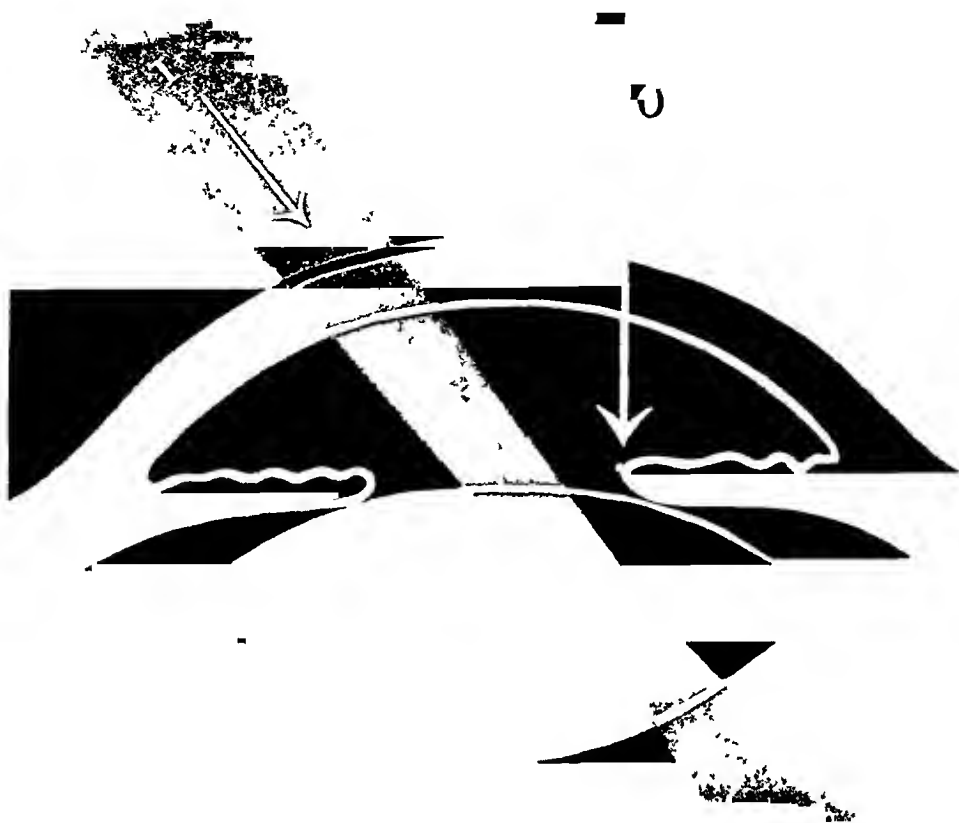


FIG. 309. Diagram illustrating the technique of retro-illumination of the iris.

layers of the highly pigmented dilator and the underlying epithelium. However, at times it is possible to see a faint radial grayish striation of the surface of the exposed pigmented layer; anatomically this represents the part of the dilator known as the posterior border lamellae. It is composed of a compressed layer of spindle-shaped processes derived from and indistinguishably connected with the deeper situated epithelial cells. However, in the lighter colored irides the sphincter can usually be seen by optic section; this is true not only of its outer edge but also of its surface, which generally has a yellowish appearance (Plate XLII).

Retro-illumination. Since it is impossible to retro-illuminate the normal iris, this method can only be used for the detection of pathologic states. The absence of retro-illumination does not exclude path-

ology of the iris but when present its significance is irrefutable.

Biomicroscopically, the iris can only be satisfactorily retro-illuminated by means of diapupillary illumination, that is, by light reflected back from the lens (Fig. 309). This is accomplished by directing the beam obliquely through the lens via the pupil. Because of the fact that there is a high degree of internal reflection and dispersion in the lens, considerable light radiates anteriorly toward the posterior iris surface. If the posterior pigmented surface is normal most of the light is absorbed. *However, when the retinal pigment is absent or has been destroyed, e.g., in atrophic states, varying degrees of this reflected light may pass through the defective areas to the eye of the observer.* Retro-illumination of an albinotic iris is strikingly beautiful (Plate XLVI, figs. 1, 2). This method is not applicable in aphakia since the vitreous is a poor reflector. Internal reflection in the lens increases with age and this is most marked when the lens is cataractous. Retro-illumination early discloses presenile or senile atrophy of the pupillary border — atrophy of the stroma and posterior pigment layers, ruptures or tears, holes, or dehiscences. Frequently cysts and efflorescences (especially at the pupillary border) can be retro-illuminated. Because of the required increased obliquity of the beam's direction as it enters the pupil, only opposite sides of the iris will be adequately retro-illuminated. For example, to retro-illuminate the temporal side of the iris, the light must be directed into the pupil from the patient's nasal side and vice versa. With experience, the observer almost intuitively looks at the darkened iris as the focused beam enters the lens and, with perhaps slight oscillatory movements of the light, will immediately note any transilluminable or rarefied areas in the iris. These will appear whitish, grayish, or yellowish (depending on the color of the light reflected back from the lens) in contrast to the darkened uninvolved adjacent iris tissue.*

Diapupillary Transillumination or Retro-illumination of the Iris in Fundus Reflex. Comparable to what occurs with the ophthalmoscope and with diascleral illumination (diaphanoscopy) it is possible under certain conditions to observe the red glow of the fundus

* To a lesser degree, the iris may be retro-illuminated by light reflected from the fundus.

reflex when the focused beam of the biomicroscope passes into the eye via the pupil (Fig. 310). When the iris is normal only the glow in the pupil will be observed. However, defects in the iris resulting

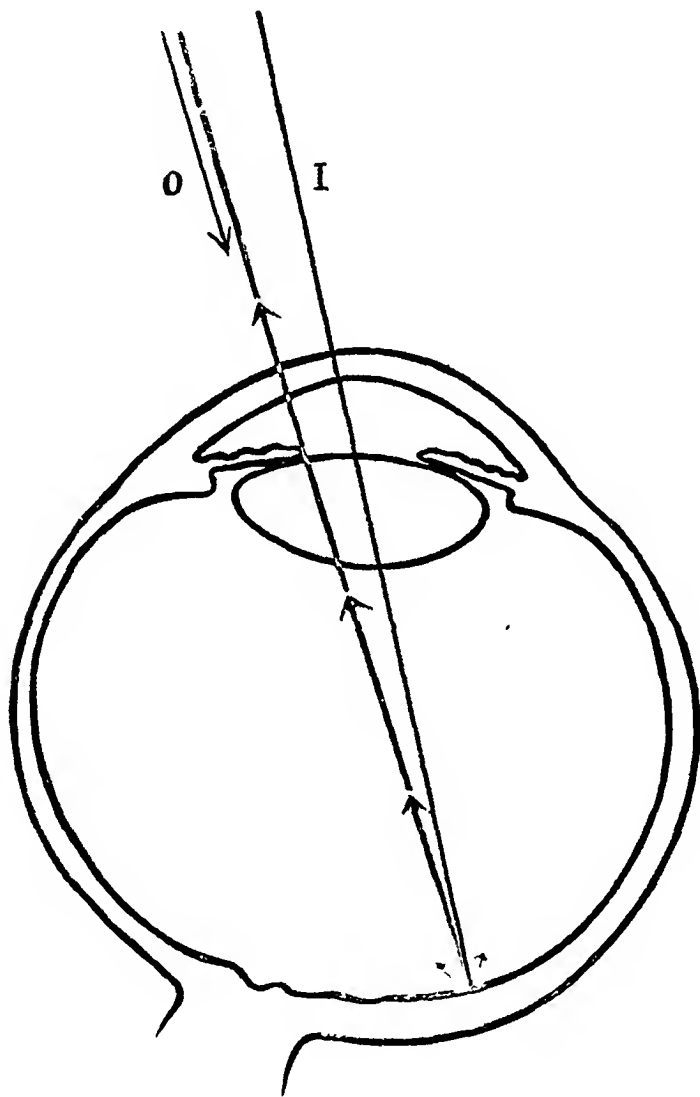


FIG. 310. Diagram illustrating the technique of diapupillary transillumination of the iris by light reflected from the fundus.

from atrophy, holes, dehiscences, and even minute depigmentations of the posterior retinal layer will permit passage of this light, which appears reddish in contrast to the grayish hue seen in ordinary retroillumination.

The direction and course of the emergent rays will vary depending on the refractive condition of the eye. In order to observe these

rays and the red glow which they cause it is necessary for the observer's direction of gaze to correspond as closely as possible to the direction of the emergent light. In other words, the head and eye of the observer must be as close as possible to the arm of the illuminating system. This corresponds to the technic of gonioscopy and biomicroscopy of the fundus. Although the employment of the Koeppel or Kleefeld mirror and a monobjective microscope assists in narrowing the angle between observation and illumination, better results are obtained, as pointed out by Davidson,³⁹³ by dispensing with the microscope and using the unaided eye. To increase the brilliance of the reflex, the 100-mm. illuminating lens is preferable to the ordinary 70-mm. In the Poser instrument* greater depth of focus can be secured by using the diaphragms. The brilliance of the fundal reflex also depends on the strength of the light source. Overloading the nitra lamp above the usual 8 volts or the use of an arc lamp is of advantage. Davidson states "the maximum intensity is secured by focusing the beam for the plane of the pupil. The beam then occupies only a small portion of the pupil, the free portion giving a fundus reflex. An overloaded voltage is necessary to secure the requisite intensity and the observer's light sense should be enhanced by dark adaptation. A small pupil is preferable in looking for defects which may disappear in the iris folds. The visibility of the iris defects seems little affected by the direction of the beam; that is, whether it is directed to the same or opposite side of the sector involved."

Proximal (Indirect) Illumination. When the focused beam falls on the iris, some of the light scatters within the neighboring stroma itself, resulting in a form of dark field illumination. For example, when the beam is focused near the pupillary edge in a lightly pigmented or blue iris and moved peripherally, ever so slightly, a portion of the crescentic outer margin of the sphincter can be observed. (See Vol. I, Figs. 99, 100.) Normally, the axial edge of the muscle is connected so intimately with the pigmented pupillary excrescences (seam) that it is not visible. The outlines of crypts, atrophic areas, and perforations can also be identified by this form of illumination.

* See Volume I, page 48.

Moreover, it is useful in the study of cysts and tumors. Frequently, following trauma, a reddish hue can be discerned in the stroma owing to the presence of bloody suggilation — a phenomenon often over-

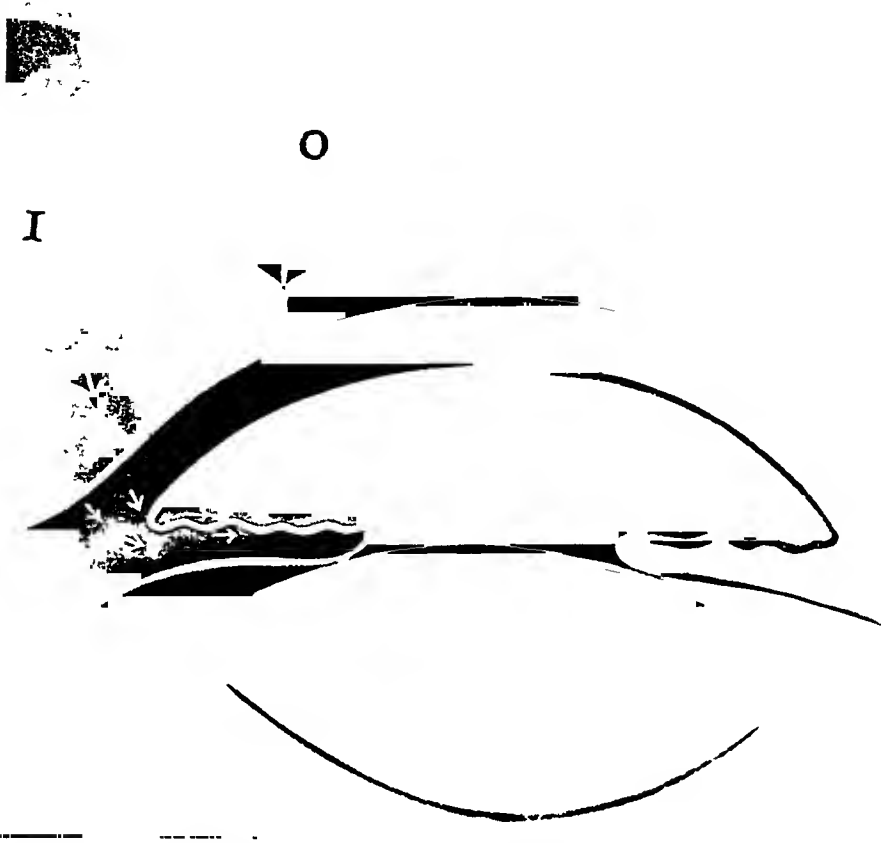


FIG. 311. Diagram illustrating the appearance of the iris when viewed by corneoscleral scatter.

looked and invisible by other methods. The incident angle of the beam should be a little wider than that used in direct focal illumination. Using the indirect method together with extremely high magnification, Koeppe has claimed that it is possible to make out in light colored irides, especially in older persons, a subendothelial system of vesicles and vacuolar lymph spaces. He described these in great detail stating that this system forms a glasslike layer having a frosted appearance. However, Koeppe's views on this subject have not been substantiated by other workers.

Iris Scatter. Transscleral and translimbal (corneal) illumination of the iris is entirely feasible providing the examining room is very dark and the light source is of high intensity. The overloaded nitra

bulb (12 to 14 volts) or better, an arc lamp, is advantageous. By slightly moving the light over the cornea a place may be found where the same and the opposite side of the iris surface (especially the periphery) are caused to glow (Fig. 311). The light is reflected from an illuminated arc of the posterior corneal surface. By this method an unsurpassed plastic impression of the configuration and molding of the iris is obtained. The iris contours, depressions, tumors, or discontinuities are displayed in relief like an itaglio. With the stronger illuminants, holes and other defects, such as are seen by diapupillary illumination, may become visible. This method corresponds to sclerotic scatter (see Vol. I) employed in the examination of the cornea. In the iris it affords a view of its entire anterior face similar to that provided by diffuse illumination. With the slit aperture wide open, in order to obtain maximum intensity of light, the beam is focused at the corneoscleral junction. The iris is then observed with the microscope, using low or middle magnifications.

Specular Reflection. Contrary to what occurs on the smooth surfaces of the cornea and lens it is not possible to obtain any zones of specular reflection in the iris. Not only does the iris absorb so much light, but the very nature of its surface precludes any sizable degree of mirror (regular) reflection which is necessary for the production of such zones. Consequently this method cannot be applied to the biomicroscopic study of the iris.

BIOMICROSCOPIC APPEARANCES OF THE NORMAL IRIS *

For our purpose the iris may be considered as being formed of two main layers (Fig. 312):

I. The anterior or mesodermal layer: composed of

- | | | |
|---|---|---|
| <p>A. Anterior
(superficial)</p> <p>B. Posterior
(deeper)</p> | } | <p>leaves, which together form the anterior face
of the iris.</p> |
|---|---|---|

II. The posterior or ectodermal layers (the posterior face) is com-

* The designs and configurations formed by the iris structures, as seen from in front, are known as the "markings of the iris."

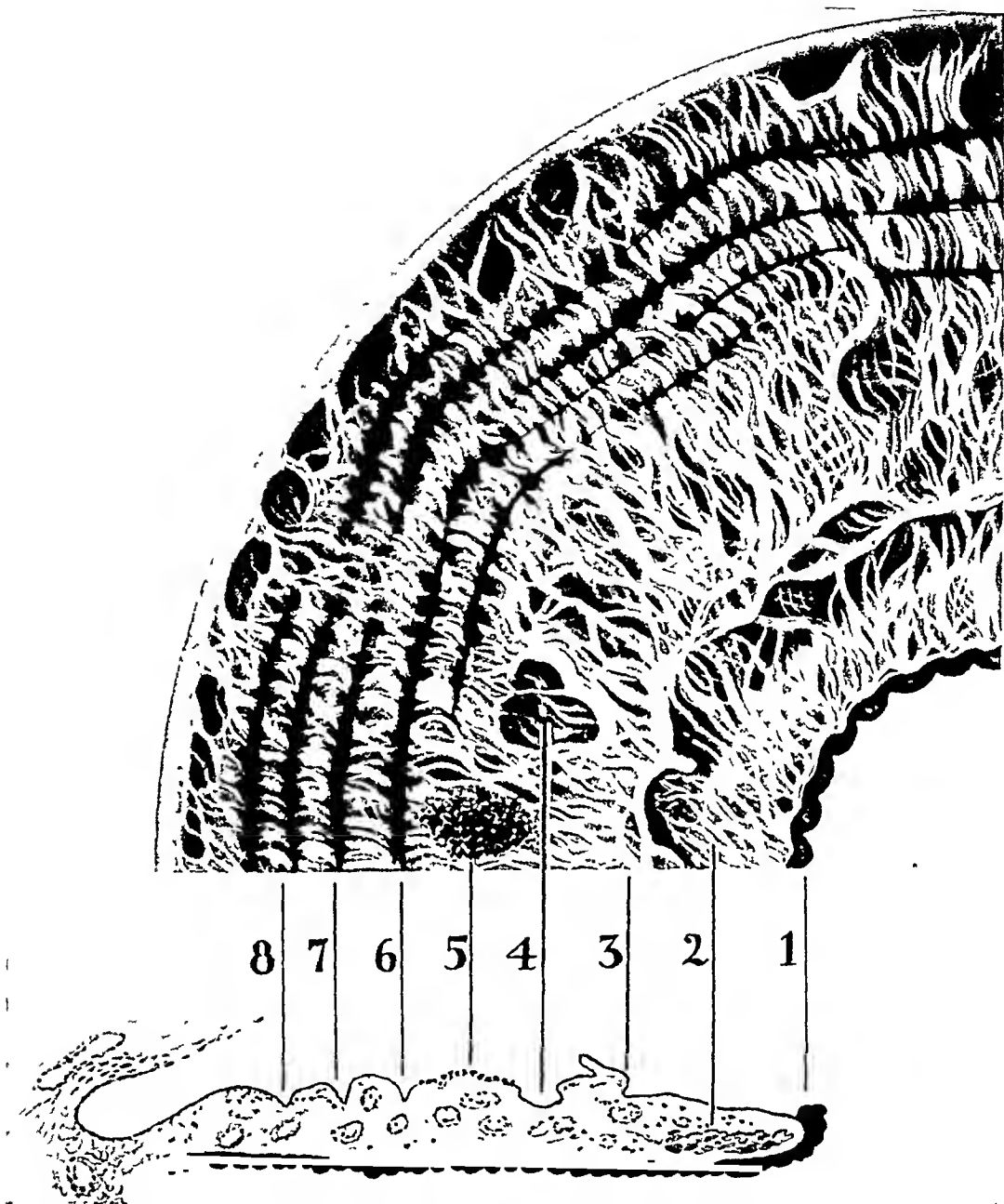


FIG. 312. Surface markings of the iris projected on its histologic section (diagrammatic). 1, Pupillary pigment border (seam); 2, sphincter area; 3, lesser circle (collarette or frill); 4, crypt; 5, freckle; 6, 7, 8, contraction furrows.

posed of dilator and posterior pigment epithelium. It is seen as one layer and can only occasionally be differentiated in normal eyes with the biomicroscope, e.g., in the bottom of a deep crypt (Plate XLIII, fig. 5).

THE ANTERIOR LAYER

The anterior layer is composed of two leaves derived from mesodermal tissue — superficial and deep. This fact was first discovered by Fuchs. Together both these layers form the iris stroma. The stroma is composed of connective tissue fibers, vessels, and pigmented cells. When a blue or gray iris is observed, it is seen to be built up of fibers, or trabeculae whose main direction is radial — converging toward the pupil. This is especially true in the ciliary region, where the trabeculae are derived from thick vascular trunks which enter the iris at regular distances.

In some cases interwoven or interlaced trabecular branches may run obliquely or even concentric to the pupillary margin. These, however, are best seen only in atrophic irides or when certain defects such as ruptures reveal the deeper layers. In blue or lightly colored irides, especially in the ciliary zone, these radial fibers have a wavy or even corkscrew appearance. They really are the blood vessels of the iris, which appear opaque and apparently bloodless because of a thick adventitia, which is enmeshed by a delicate veiling of stromal tissue, resembling an insulated electric wire. Overdevelopment of the anterior leaf in the region of the lesser circle may obscure the striation. Except in the region of the lesser circle, where a number of larger trabeculae condense (frill) and almost turn at right angles to run concentrically, trabeculae tend to run more or less parallel in a radial manner. With higher powers it will be seen that they frequently branch dichotomously and at times give off branches which cross neighboring trabeculae and run obliquely or even somewhat concentrically for a short distance. Such branchings are best observed in the ciliary region and near the pupillary zone. Occasionally, in blue or gray irides, by using a blue filter and strong direct light, one can make out a dark central channel in a larger trabecula.

PLATE XLIII

FIG. 1. Marked senile atrophy of the pupillary border.

FIG. 2. Displaced pigment excrescence on the surface of the iris below.

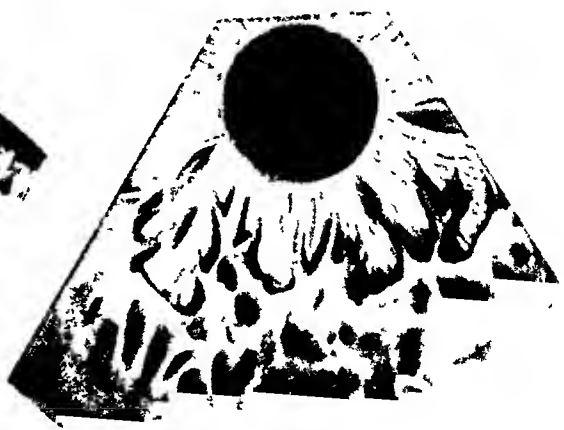
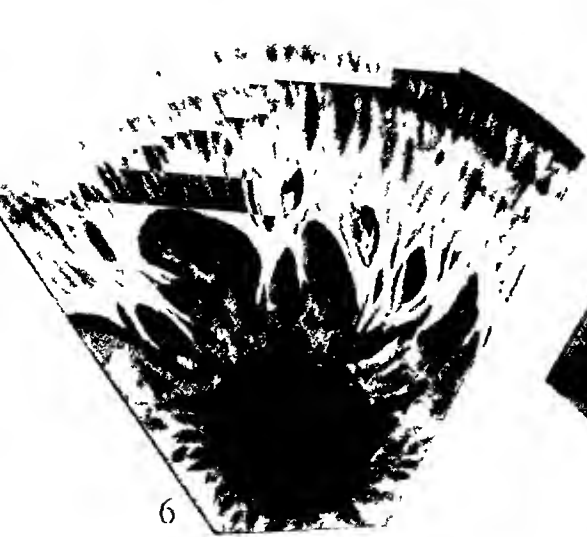
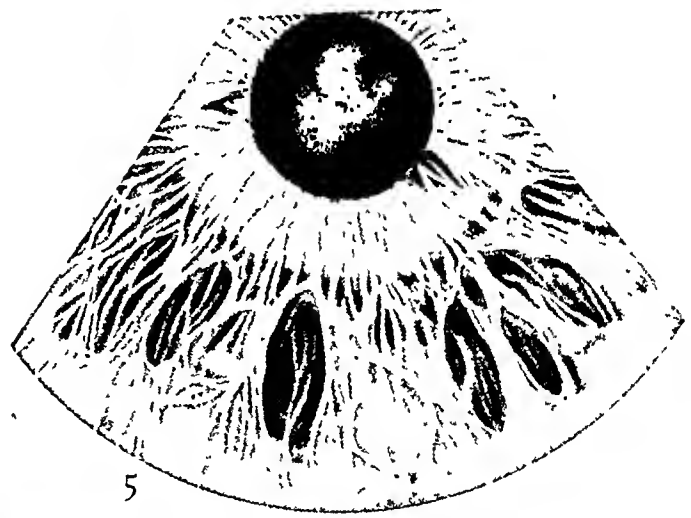
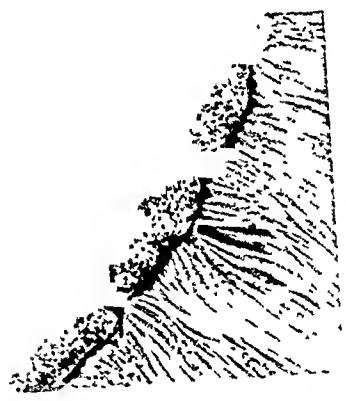
FIG. 3. Senile atrophy of the deeper leaf of the iris with marked rarefaction of the stroma.

FIG. 4. Senile hypertrophy of the pigment excrescences of the pigment border.

FIG. 5. Senile rarefaction and atrophy of the pupillary iris border. Note sphincter.

FIG. 6. Iris bicolor. Ciliary portion of the iris is gray while the pupillary portion is brown.

FIG. 7. Iris bicolor. Ciliary portion of the iris is brown while the pupillary portion is gray.



This probably is the blood column.* If the stroma is very light, occasionally one may, in addition, see a fine pink vessel traversing the trabeculae (Plate XLI, fig. 6). These vessels are enmeshed in the soft cottonlike tissue of the stroma and usually are found in the region of the lesser circle and in the periphery. However, one is less likely to see them when the trabeculae of the superficial leaf are overdeveloped.

Biomicroscopically the visibility of these structures will vary from case to case, depending on the design or compactness of the tissue and on the amount of pigmentation present. As a rule the light colored iris has a more delicate and fragile architecture than the dark (Plates XLI; XLII). The structure of the anterior layers of the gray iris is frequently looser than that of the blue and consequently may allow of better inspection of the deeper leaf. In some blue irides the superficial layer of radial fibers are very compact, being without crypts or other sharp lines of demarcation, such as, frills or contraction furrows. Brown irides may also display varieties of structure compactness. In some, a marked and beautiful design of trabecular structure may be seen in which the frill and superficial trabeculae stand out sharply, even as strong thick bands (Plate XLII, fig. 5). Some of the larger trabecular bundles may subdivide into smaller strands. The finest strands or threads are those which cross over or subdivide the crypts. However, the surface pigmentation and fiber structure may be so compact as to give the dark brown iris a uniform velvety or spongy appearance to a degree that no surface markings may be visible (nontrabecular construction) (Plate XLII, fig. 1). It is not possible biomicroscopically, even with the highest power, to discern any structure on the iris surface, which might be interpreted as an endothelial cover. With the optic section the anterior border layer is discerned as a thin solid line.

* Koeppe described the iris trabeculae in great detail, according to their size, branchings, and as to whether or not they had visible vessels. He was able to discern many more vessels bearing trabeculae than I have. My impression is that in light colored irides, especially in the aged, more actual vessel-bearing trabeculae can be seen than in the young but on the whole they are only occasional. When using the highest power, i.e., over 70 X, great enlargement of the structures is obtained but at the expense of clarity, since even with the brightest light the enlarged images appear blurred.

In light blue irides with 40 \times magnification or more, using indirect illumination, I have been able to make out what appears to be a fine stippling in the delicate cottonlike connective tissue supporting the superficial trabeculae. But even with still higher magnification I have not been able to convince myself that these were actual vesicles such as Koeppe described.

The superficial leaf of the anterior layer of the stroma starts at the periphery and extends to the lesser circle (circulus minor), ending in an irregular zigzag line or frill that runs irregularly concentric to the pupil. This line may be found at varying distances from the pupillary margin but usually closer to the sphincter edge than to the periphery. Although it frequently appears as a single line, in reality it consists of many bowed trabeculae bound together by connective tissue. The superficial leaf is usually so intimately connected with the underlying deeper leaf that except in certain types of irides it is not possible to distinguish it as a separate layer.* But at its termination at the lesser circle it may be separated from the deeper leaf by a frontal cleft, known as "Fuchs' cleft." This cleft can easily be seen in the partially dilated pupil where the pupillary zone begins to slide under the frill, which stands out as a more or less sharp ridge (Fig. 313 B). In most cases there are direct connections between the frill and the radiating fibers of the deeper leaf of the pupillary zone. These trabeculae extend to the pigment border of the pupillary margin. With higher magnification it will be seen that although most of these trabeculae run axially to the pupillary margin, dichotomous branchings often occur in which some of the smaller branches tend to meet divisions from other trabeculae; in so doing they take a course somewhat concentric to the pupil. Near the pupillary margin an occasional trabecula may loop backward to join another radial branch.

It should be remembered that embryologically the superficial leaf is composed of radial blood vessels, supported by connective tissue,

* Congenital splitting of these two layers has been reported by Waardenburg. It was transmitted from a mother to three children as a dominant hereditary characteristic. Such a change can also occur as a senile degeneration. (See page 777. Also see Plate III, fig. 5.)

which form circular arcades in the region of the lesser circle. The convexity of the arcades faces the pupil (Fig. 313). From this circle terminal arcades of vessels, also supported by a delicate film of mesoderm, convergently extend over the pupillary opening to form

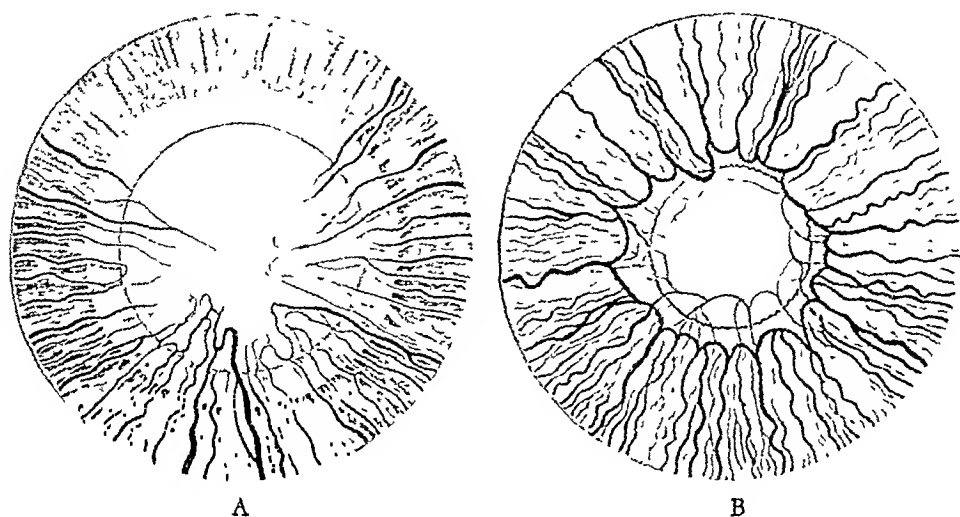


FIG. 313. Congenital vascular arcades in the region of pupil. (After Ida Mann.)

the pupillary membrane. Hence, the frequently seen postnatal remnants of the pupillary membrane are always connected (at least at one end) with the frill. (See Vol I, Plate XXXVIII, figs. 1, 2, 3, 4, 6.) From the sixth month of fetal life the vessels of the pupillary membrane no longer grow but instead begin to atrophy and finally disappear, at the same time the vessel arcade of the lesser circle continues to develop with the remainder of the iris. The vessels of this arcade have a greater lumen than the intervening or deeper vessels, and later they appear of heavier structure. Hence, the frill may be considered a form of redundant hypertrophic rudiment of the fetal iris vessels of the lesser circle; consequently the architecture of the frill and the superficial leaf repeats the fetal vascular design. Biomicroscopically, the appearance of this zone varies so much from one case to another that it is impossible to give any systematic description of it. At times, even in adults, with strong illumination (arc source) blood in the form of fragmented columns may be seen in certain parts of the frill itself. That the superficial leaf is really a definite layer is proven by the fact that it may be missing,

or if present may become detached, in which case it has even been seen to float freely in the anterior chamber.

In cases in which no frill is found it must be assumed that resorption of the vessels was complete and that the secondary hypertrophy of the vascular remains at this point did not occur. In others the frill may be so interwoven with neighboring fibers as to be invisible or, frequently, strands flush with the surface may lose themselves in the direction of the pupil (Plate XLI, fig. 9). The frill in certain instances (e.g., in lighter colored irides) may be so well developed as to constitute a hypertrophy. Vogt described such a case in which there was also a thick and strong extension similar to a pupillary membrane. These fibers are very expansive and do not tear with mydriasis. In such cases a weblike structure frequently is seen bridging over the angles formed by the junction or anastomoses of the trabeculae. This, together with fusion of the trabeculae into bundles, may give these structures at the lesser circle an appearance of giant size (Plate XLII, fig. 5). Here they may flatten and spread out fanwise or appear like stars.

There are cases in which the frill extends over to or is attached to the pigment border of the pupillary margin. In others, it may be found near the chamber angle. Frequently, it happens that the frill is only well developed for a small distance, and in the remaining iris circumference only an unmarked raised area indicates the beginning of the sloped pupillary zone.

Krückmann-Wöfflin bodies. At times nodular structures may form in the region of the collarette. More commonly similar knob-like condensations (brown, yellowish, or whitish in color) may be found at the periphery near the limbus (Plate XLII, fig. 5). These nodelike structures lie between the contraction furrows and with mydriasis they assume a concentric arrangement owing to deepening of the furrows. Deepening of the contraction furrows also tends to exaggerate their height so that they resemble mounds bordered by troughs. The color of these nodes vary with iris coloration. In very dark brown irides they appear darker since condensations of the superficial iris layer results in a localized heaping together of the

melanophores (similar to nevi) while in irides of lighter color, owing to the sparseness of pigment tissue, these condensations appear yellow or even whitish. They are best seen by indirect light and when present usually appear in both eyes. In cases of iridocyclitis they have been incorrectly interpreted as inflammatory nodules. Comparison with the unaffected eye will serve to avoid such an error. These normal tissue nodules were described by Krückmann and Wöflin and are best seen by iris scatter or indirect illumination.

Crypts (holes or openings) in the stroma are usually found in the superficial leaf near the region of the frill; however, they may occur in the far periphery (Fuchs) or in the deeper leaf of the pupillary zone (Plate XLII, figs. 4, 5, 7). Crypts probably result either from defects or from localized areas of fetal or postfetal atrophy, involving not only the superficial leaf but also the deep one as well. They are only found in man. Even other higher mammals, such as monkeys, do not show them. Crypts are seen in both blue and brown irides but in the typical spongy type of Negro iris, if present, they are chiefly found in the pupillary zone. Crypts of the peripheral ciliary zone are usually smaller and shallower than those of the pupillary area or in the region of the frill.

When the structure of the deeper leaf is seen in the bottom of a crypt it appears lighter in color in direct focal illumination than the neighboring surface iris which normally bears most of the stromal pigment; in deep crypts and especially in those which extend entirely through the posterior leaf, the dark posterior retinal pigment layer is seen. By daylight, however, a crypt may appear darker than the surrounding tissue owing to the fact that its base lies in the shadow cast by its steep side walls. This darkening effect will also be augmented by the darker color of the posterior surface of the iris when the depth of the crypt extends down to it. Occasionally in large open crypts trabeculae bearing small radiating vessels containing blood may be seen in the depths. Crypts may be subdivided by thick or thin tissue threads passing over their openings. This may also occur within the crypt giving it a layered appearance. The walls of a crypt show the same structure as the stroma. The larger

crypts are rhomboidal in shape and are usually bordered by large trabeculae. With dilation of the pupil these tend to flatten out and take a direction concentric to the pupil. The smaller crypts have a more circular opening. In light colored irides Koeppe described crypts of the third magnitude, i.e., crypts so small that they could be seen only with magnifications of 100 \times .

The anterior border layer is usually entirely missing over the openings of crypts and the deeper stroma itself, seen in the depths, is less pigmented. However, in some cases, as previously pointed out, an occasional trabecula may pass over the opening or within the depths of a crypt. Such fibers vary in size and at times are merely threadlike. Except for this, even after employing the narrowest possible optic section and high power, I have never been able to find any covering over crypt openings that might be interpreted as an endothelial or anterior border layer.

Contraction furrows are seen in the surface of the iris peripherally to the region corresponding to the middle half of the ciliary zone (Plate XLI, fig. 1). These are sharply outlined grooves, running concentric to the peripheral iris border and probably result from pupillary movements. An individual furrow may extend a quarter or half way around the iris but it never completely encircles the iris (Fig. 312). Similar to the lines on the palms of the hands, which deepen when the hand is flexed and flatten out with extension, the furrows deepen with mydriasis and flatten out with miosis. As is the case in crypts, the troughs of the furrows in a brown iris, especially in children, are lighter in color than the surrounding areas. This may be caused by the fact that anatomically the anterior border layer is thinner in the bottom of the furrow, hence there is less pigment. Mawas and Krückmann both believe that the lessened pigmentation results from mechanical changes, while Vogt explains this as an anomaly of development. In very light colored irides, owing to the paucity of stromal pigment, the bottom of the furrows have a dark appearance. Fine radial strands of the superficial leaf may be seen in the furrows of heavily pigmented irides.

The posterior or deep leaf of the anterior layer is covered by the superficial leaf, from its periphery to the frill (ciliary portion of iris), from here to the pupillary margin the stroma is seen to be formed mainly by the deeper leaf and lies exposed to view. When not too compactly obscured by pigment, this deep stroma can be observed as thin, stretched trabeculae, running radially and at times slightly eccentrically (Plate XLII, figs. 5, 6, 7). As stated before it is not always possible to identify circular fibers (i.e., those concentric to the pupil): occasionally, particularly in the normal blue or gray iris, radial vessels may be seen in this area. In miosis, in cases in which posterior synechiae exist, or following iris atrophy, trabeculae of the deep leaf may be stretched tautly between the frill and the pupillary margin. However, the fibers of the adjoining superficial leaf may reveal no such tension — remaining wavy and relaxed, indicating the independence of this leaf to changes occurring beneath it. In other words, the superficial leaf will only act passively with the movements of the deeper one, which is more intimately connected with the sphincter and dilator, structurally and functionally.

In the ciliary zone the structure of the deeper leaf can be seen only through openings or crypts in the superficial leaf. When the structure of the deeper leaf is visible, it is seen to be composed of fine, compact, gelatin-like, sometimes branching, radial fibers, frequently unlike those of the superficial leaf (trabecular form); the latter are apt to be composed of thicker, whiter and more loosely formed strands that tend to be wavy, and to anastomose and at times form arcades.

The Pigment Seam (Excrescences) of the Pupillary Border. Seen from in front, the normal pupillary border of the iris (pupillary termination of the deeper leaf) is marked by the presence of a black-brown seam which, when magnified, appears beaded, especially when the iris is not dilated (Plate XLI, figs. 1, 2). However, only the side toward the black pupil has this appearance; the stromal edge is smoother. By daylight this seam appears almost black, but with the sharply focused beam of the biomicroscope it is always dark

brown in color. As the pupil dilates and the pupillary zone of the iris draws back under the frill and ciliary portion, the pigment seam loses its beaded character and becomes drawn out in the form of

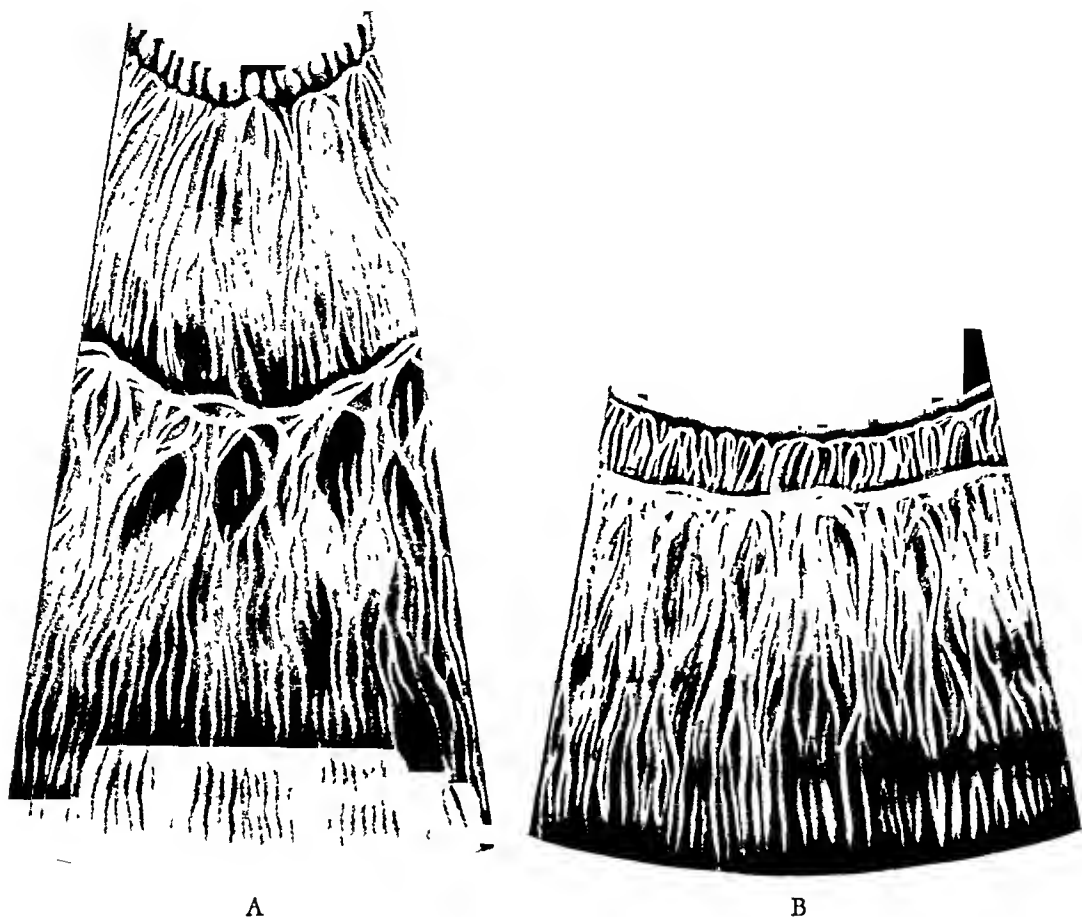


FIG. 314. A. Surface markings of iris (contracted pupil). B. Appearance of surface markings of iris with pupil in mid-dilation.

a thin line (Fig. 314 B). Hence, it is always seen better in the undilated pupil (Fig. 314 A). After miosis, a previously almost invisible pigment seam may become prominent (Plate XLIV, fig. 6). When examining the border with the focused beam, it is advisable to have the beam directed from the opposite side in order to see the curved edge which is directed toward the lens. With occasional exceptions, the average width of the pigmented pupillary seam varies from 0.04 to 0.1 mm. when the pupillary diameter is about 3.5 mm. In Negroes it is somewhat wider. In children, because the pupil is wider normally, the pigment border is narrower and is not seen as well as in adults. In a series of 63 children between the ages of 9 and

10 Fuchs found that when the pupil had an average size of from 4 to 6 mm., the upper pigment border is .05 mm. in width. If the pupil is narrowed to 1 mm. a widening of the pigment border occurs up to 0.1 mm. or more.

Individually the beads or prominences (excrescences) of the seam vary in size and are curved toward the lens. They are separated one from the other by crevices which change with pupillary movement. The crevices are not always equidistant from one another. At times, with higher power, it may be seen that tongues of pigment extend peripherally from the base of the excrescence to the stroma. In other cases the attachment to the stroma is smooth and sharp but generally one gets the impression that the pigment edge is slightly rolled in. With higher power it will be seen that the surface of the excrescences, especially near the pupillary side, is not smooth but is granular owing to the deposition of small dots of pigment. Near the pigment excrescences and lying over the area occupied by the sphincter, small globes of retinal pigment may occur either singly or in groups. When large they may resemble nevi. These probably represent the clump cells of Koganei (Salzmann).⁵⁹¹

Normally, the upper part of the pigment seam is generally wider and more dense than the lower. By micrometry Vogt observed this difference in more than 50 per cent of cases examined. Although the comparative width of the upper and lower seam is variable, in a series of cases described by Vogt the upper border measured between 0.05 and 0.06 mm., and the lower from 0.01 to .02 mm. in children from 6 to 16 years, while in adults it was 0.065 mm. above and 0.03 to 0.04 mm. below. In rare instances it may be twice to three times as wide above as below. Frequently the beading effect of the pigment of the lower pupillary border is more distinct even though this border is narrower. Occasionally the pigment excrescences may be missing in this region. This has also been found in association with the so-called physiologic hypoplasia or partial coloboma of the mesoderm, commonly occurring in the inferior segment of normal blue irides (Plate XLI, fig. 4). Considering the fact that this type of hypoplasia occurs in a normal iris and that the missing

pigment is not found deposited elsewhere (e.g., on the stroma as might occur with certain anomalies or in disease) such a phenomenon cannot be considered as pathologic.

Total absence of the pigment excrescences is rare. Koby has described a case in which this was found. Before making such a diagnosis, as Mawas has pointed out, the pupil should be examined in miosis. The difference in width between the upper and lower pigment seams was first noted by Fuchs, who stressed the fact that this variation was not caused by any tendency to entropion below and also that it may occur unilaterally. One may be called upon to differentiate such a hypoplasia from an acquired defect. In the former the decrease in width is gradual and extends over a considerable area, while in the latter it tends to be sharply localized. The tendency to overdevelopment of the upper pigmented border may likewise be genetically determined — a condition whose extreme state is seen in flocculus (Plate XLIV, figs. 4, 5). Some investigators have even gone so far as to consider this condition atavistic, since overdevelopment of the upper pupillary pigment border is normal in horses and ruminants.*

Infrequently, an irregularity of the pigment beading occurs in which the beading is slightly angulated or dentate (Plate XLIII, fig. 4). This may be due to invisible defects in the stroma which results in less traction at these points.

The Sphincter Iridis. Biomicroscopically the sphincter of the normal iris can be seen only in occasional cases and then only in blue, gray, or lightly pigmented irides. Lying deep in the stroma of the posterior leaf of the pupillary zone, the visibility of the sphincter depends not only on the pigmentation but also to a greater degree on the density of the overlying trabeculae (Fig. 312). Hence in some cases only the distal edge of the sphincter can be faintly discerned, while in others its full width is apparent (Plate XLII, figs. 2, 3). In the latter case it is seen as a grayish or more commonly as a yellowish feltlike band, concentric with the pigment seam.

* In certain animals hypertrophy of the border pigment (flocculus) may be extreme; this is thought to be a protective defense against light coming from above.

This band measures from 0.7 to 1 mm. in width, varying with the size of the pupil. In structure it seems to have a uniformly solid consistency and even when highly exposed, e.g., in albinism, senile rarefaction, atrophy of the iris, or in blue irides of light texture, it is impossible to make out any details or striations in its structure (Plates XLIII, figs. 4, 5; XLVI, figs. 1, 2). Although frequently discernible in diffuse illumination, the sphincter is best seen by reflected light as obtained by indirect illumination. Oscillatory movements of the beam will display its high degree of mobility. Depending on the amount of dilation, the width of the sphincter may narrow by 0.2 to 0.4 mm. In maximal dilation it becomes invisible owing to the fact that the entire pupillary zone telescopes under the ciliary part of the iris.

IRIS COLORATION

The color of the normal iris is determined by genetic factors. The working of these factors is as yet not completely understood, though the following facts are found to be true in the majority of cases: Blue color is recessive to gray, green, and light brown, which in turn are recessive to dark brown. A person who has dark brown eyes (a color transmitted as a Mendelian dominant) has, therefore, at least one parent whose eyes are dark brown, while on the other hand a child of parents who both have dark brown eyes may be light-eyed. Since, however, the darker colors are not always fully dominant over the lighter ones, mixed colors may appear occasionally instead of pure dark or pure light colors. Then again, although blue eyes theoretically are homozygous, all the children of two blue-eyed parents should be blue-eyed. Actually this occurs in only 88 per cent of cases. Twelve per cent of the cases have brown eyes, showing that although both parents had blue eyes, at least one of them must have had a "latent" gene for brown.

Anatomically, the color of an individual iris depends on the presence or absence of stromal pigmentation (melanophores) and on the density and thickness of the stroma. Except in albinism the posterior retinal pigment layer is always dark brown in color. When

the stroma is unpigmented, its tissue is white or gray. By diffraction, the passage of light through this tissue gives the impression of blue or gray depending on the compactness and translucency of the unpigmented stromal tissue. When the tissue is less transparent, the iris is gray. A similar phenomenon also determines the blue color of the cerulean lens opacities, in which thin whitish opalescent opacities are viewed against the background of the black pupil. The blue color of the sky can be explained in the same manner, i.e., diffraction of light occurs through the atmospheric haze against the darkness of the heavens. The presence of melanophores in the stroma gives the iris its brown color.* These cells, when present, are most prevalent in the anterior border of the iris although the deeper trabeculae, when visible, will also be seen to be pigmented. Contrary to what occurs in the fundus, in the iris, owing to the thickness of the adventitia of its vessels, practically no reddish reflex results from its illumination and hence the rich vascularization of the normal iris tissue plays no role in its coloration. When the stromal pigmentation is sparse, the iris color may vary from a greenish to hazel color (Plate XLII, figs. 6, 7, 8). However, when the stroma is uniformly pigmented the iris will be brown. When the stromal pigmentation is greater, the iris appears darker brown. Even in the Negro, whose iris (by low degrees of diffuse light) may appear black, actually in direct focal light it is always dark brown. This is also true for the color of the pupillary excrescences. One must always use caution in judging the exact color of tissue or cells when using the biomicroscope. Unless monochromatic light is employed, apparent differences in color will be confusing. For example, an iris that appears to have a brilliant light-blue background by daylight may have a light brown background by focal illumination (Plate XLI, figs. 7, 8). In each case the surface trabecular fibers will be whitish. In other words, with the focal beam, sufficient brownish or yellow reflection from the pigmented posterior face of a blue iris may give it a brownish or yellowish tinge.

* In certain albinos the iris, although devoid of retinal pigment, may have a brownish color if stromal melanophores are present.

The varieties of normal iris coloration are so different from individual to individual as to defy any systematic description. In a sense it is difficult to separate variations in color from those of structure since the visibility of the iris architecture (trabeculae and surface markings) depends on the degree of pigmentation. In the main, two types appear: (1) light colored irides (blue or gray) having a visible fibrillary structure and (2) brown (light or dark) irides which may reveal a trabecular structure or may have a uniform smooth or spongy surface devoid of any trabecular architecture. However, in both types of irides the mesodermal layer can exist either in rudimentary form or in a state of full development. In the latter case the frill (collarette) and the superficial crypts and trabecular arcades will be present.

Ordinarily in brown irides it is impossible to discern individual melanophores. However, with high magnification in the smooth-surfaced dark brown iris (without trabecular structure) one receives the impression of a fine surface net of delicately branching starlike structures.

Isolated spots or groups of pigment spots (freckles) varying in size and shape, are commonly seen in irides of all colors and are found in all parts of the iris surface (Fig. 312; Plate XLII, fig. 6). With the slit lamp it is possible to observe the physiologic flat spots by indirect illumination, i.e., focusing the beam alongside the spot so that some of the reflected light actually retro-illuminates them. Using this type of illumination it will be seen that the spots are granular in character and are made up of irregularly sized pigment dots. Even though they appear solid to the unaided eye, it is the author's belief that the granular aspect of physiologic iris spots differentiates them from nevi or melanomas; the latter are more solid and closer in appearance to the ectodermal pigment excrescences of the pupillary margin. Frequently some of the pigment grains outline a trabecula on which they lie. By direct focal illumination with the narrow beam one gets the impression that they lie directly on the iris surface and not within the tissue itself. Occasionally, when the spot is near the pupillary margin, it may send proc-

esses (pigmented) to the seam. But these will always show a difference in color and structure from the retinal pigment of the seam.

In blue irides multiple pigmented spots may result in the so-called "piebald iris," in which dark brown patches distributed around the circumference of the iris stand out in bold relief (Plate XLII, fig. 7). This type of spot is seen even in dark irides of uniform surface pigmentation; It is thought that those situated near the sphincter are formed from the clump cells of Koganei, which embryologically appear to be derived from the ectodermal pigment layer. When numerous or extensive, such areas of pigmentation, especially if more marked in one eye, can result in a picture resembling a partial physiologic heterochromia, and in the extreme state, even melanosis iridis (page 806). Whether or not it is proper to consider "nevi" as being derived from such physiologic iris spots brings up the conjectural question of the origin of pigment and the derivation of pigmented tumors. At the present time it would seem that the physiologic iris spots are formed from condensation of melanophores; if it were proved that melanophores were derived from the neuro-epithelium then it might be correct to postulate that nevi could be formed from the massing or proliferation of physiologic iris spots. But there is considerable evidence to show that mesodermal tissue has the function of melanogenesis; hence we shall consider that the physiologic iris pigment spots differ from nevi, at least in so far as their origin is concerned.

The use of the term "nevi" to describe the more tumorlike masses of pigment spots on the iris surface or in the stroma is unfortunate since they do not contain the typical nevus cells found in cutaneous nevi. In the case of the iris they are merely a concentration of the usual benign melanophores in normal iris tissue. Although cases have been reported in which these spots took on a malignant transformation (melanosarcoma), it is still doubtful whether this is actually so. Fuchs (1917)⁴³⁹ believed that the pigment spots previously seen at the site of a developing malignant iris melanoma were not benign nevi but actually slow growing malignant cells. It must be admitted that, when they are very small, it is not

always possible with the biomicroscope to differentiate between these "physiologic" pigment spots (nevi) and those of beginning malignancy.* However, the ordinary "nevus" found in irides of different colors are generally composed of groups of brown pigmented cells in an area of condensed normal tissue of the anterior border layer of the iris (which favors a mesodermic origin). According to Streig (1915)⁶²³ these are only transition structures or the expression of partial heterochromia. The margins of these cell groups are not sharp but merge irregularly into the neighboring stroma. Histologically the pigmented processes of these cells composing the nevus often reach into the deeper layers of the stroma (Fuchs, 1913).^{437, 438} Similar nonpigmented structures in the iris are the grayish Wolffian bodies or nodules frequently seen at the periphery of normal irides.

One should make a differentiation between the so-called pigmented nevus and the benign melanoma. The term "melanoma" should be reserved for those structures in which a small hyperplasia of the posterior ectodermal (retinal) layers occurs. Such a benign epithelial growth may occur at the pupillary margin (as in flocculi) or over the pillar edges of a coloboma, or it may be seen protruding anteriorly through the stroma, appearing as a dark spot or mass. In the latter case the site of predilection is near the sphincter edge (region of the clump cells, Koganei) but no area of the iris is exempt. This type of growth at the pupillary margin may be solid or cystic. However, when its protruding surface is small and flat, it is usually impossible to differentiate a benign melanoma from a nevus. Not infrequently this form of proliferation so resembles a malignant growth clinically as to be indistinguishable from it. Any increase in growth of one of these melanomatous spots should be regarded with suspicion. In a case of neurofibromatosis (von Recklinghausen's disease), Goldstein and Wexler (1930) found well-formed melanomas of the iris associated with nevi iridis. Other

* Even using higher powers, one cannot discern any delicate branching processes of the pigment cells characteristic of melanophores. If this were possible, it might be a means of distinguishing between the harmless nevi and malignant melanomas.

authors have described small pinpoint dots of pigment; Vogt has reported ectropion of the retinal pigment.

A commonly encountered form of iris coloration known as double coloration is that in which the color of the anterior leaf (from the periphery to the collarette, i.e., the ciliary zone) differs from that of the visible deeper leaf (from the collarette to pupillary border, i.e., the pupillary zone). One or the other of these parts in an individual iris may be pigmented or light colored, for example, the ciliary part may be brown and the pupillary part light or vice versa (Plate XLII, fig. 8). This, naturally, may affect the visibility of the delicate fibrillar stromal structure in the pigmented parts, since, as previously mentioned, they can only be seen in light colored irides or the unpigmented parts of a mixed colored iris.

Simple heterochromia (heterochromia iridis) seems to be a hereditary condition characterized by a definite difference in color between the two eyes or by zones of different color in a single iris. (This is in contrast with complicated heterochromia in which the differences in color can be explained by the presence of some pathologic process.) Simple heterochromia can be total or partial (heterochromia iridum). Several geneticists have compiled pedigrees proving the hereditary transmission of this anomaly; it is more common among animals than in man.

According to Kraupa (1923), color variations of the iris may be classified as follows: ⁵¹¹

- I. Primary — congenital or dependent on a congenital substratum.
 - A. 1. Uncomplicated heterochromia
 2. Melanosis iridis, especially with star formation
 3. Double coloration in a single iris
 - a. Iris bicolor (blue green with yellow mesopigment)
 - b. Nevus iridis (partial melanosis)
 - c. Ectodermal pigmented deposition.
 - (1) Clump cells
 - (2) Ectropion uveae
- B. Heterochromia (E. Fuchs)

II. Secondary (neurogenic)

A. Sympathetico-heterochromia

B. Following intra-ocular disease (iritis, glaucoma, hemorrhage, siderosis)

SIZE AND MOBILITY OF THE PUPIL

Ordinarily the pupillary diameter of the normal iris varies in size from 3 to 5 mm. according to differences in illumination and with accommodation and convergence. As a rule, it is wider (1) in children than in adults, (2) in females than in males, and (3) in myopia than in hyperopia. Biomicroscopy affords the best method of observing pupillary mobility. Because of the usual oscillatory movements of the eye while being examined with the focal beam, varying amounts of light enter the pupil, resulting in almost continuous rhythmic dilatations and contractions. The trabeculae of the deeper leaf in the pupillary zone are intimately connected with the sphincter and instantly respond to its movements. As the pupil dilates, these fibers become shorter by wrinkling and similarly lengthen and straighten out during contraction. In a passive way the fibers of the superficial leaf indolently follow these movements.

When the fibers of the pupillary zone run in an eccentric direction toward the pigment margin, frequently crossing others, action of the sphincter results in a scissorlike movement of these fibers. In the early stage of mydriasis, as the pupil begins to widen, the pigment excrescences of the seam begin to stretch out and to lose their beaded character. Eventually, the seam is drawn out into a thin fine line and with maximal dilatation may be hardly visible (Fig. 314 A, B). At the same time the pupillary zone slides under the frill and superficial leaf. Before the pupillary zone disappears, the cleft of Fuchs becomes exaggerated; finally the frill itself seems to form the pupillary margin. As dilatation continues, the remainder of the visible iris (now including the invisible telescoped pupillary zone) contracts itself as a unit. The contraction furrows deepen in a way similar to the action of an accordion, thus accommodating to the thickening of the tissue, which must fold in order to occupy a smaller area. As

mydriasis wears off, the same phenomenon occurs in reverse. The contraction furrows flatten out; then the pupillary portion becomes visible, sliding axially from under the ciliary part. As the pupil becomes smaller, the seam widens and the crenations or beadings are more marked. In miosis the puckering of this pigment border (which is firmly connected to the axial edge of the sphincter) may actually appear as if it were somewhat everted (physiologic ectropion) under the purse-string action of the sphincter. When this occurs, the optic section will reveal a fortuitous widening of the space between the iris pigment border and the anterior lens capsule, enhancing the communication between the anterior and posterior chambers. It is interesting to observe the action of the trabeculae during contraction and dilation of the pupil. This can only be seen in lightly colored irides where the movements of the individual fibers can be followed. The greatest action is seen in the pupillary zone and in the region of the lesser circle. The amount of visible movement varies inversely as the ciliary region is approached. When the pupil contracts, the angles formed between the crossing and branching trabeculae narrow (the movement reminding one of the opening and closing of innumerable pairs of scissors) while during dilation these angles widen.

The pupillary opening is not situated in the exact center of the iris diaphragm; but a little to the nasal side and below the center. According to Koeppe, this results in a slight rarefaction of the trabecular system in the middle temporal sector and is apparent in the light colored irides of the young. This physiologic decentration of the pupil probably is related to the embryologic closure of the fetal cleft. The temporal rarefaction together with the similar defect in the lower part of light blue irides, previously mentioned, does not affect the circular shape of the pupil. It is still unknown whether or not the iris trabeculae are elastic or whether they exert any traction effect in the mechanism of pupillary motility. The normal circular shape of the pupil may rather depend on the oblique connections between the sphincter and the dilator (page 733). These connections may serve to flatten and extend the sphincter. However,

it must be understood that pathologic variations in stroma can affect the shape of the pupil, in addition to other causes.

THE SENILE IRIS

As part of the aging phenomena the iris undergoes progressive changes. There is no doubt that the tendency of tissues to show the signs of senescence varies with the individual. In some persons, progressive changes such as arcus lipoides, iris atrophy, cataracts, vitreous degeneration, and graying of the hair, appear relatively early (presenile changes), probably induced by hereditary determinants, similar to the abiotrophies. The factor of nutrition must also be kept in mind. In others, these changes may occur either in great age or not at all. There is certainly no uniformity in their appearance as indicated by their selective presence in one tissue and absence in another. It should be remembered that changes identical with those of senility can occur in the young after disease. The type of change visible by means of biomicroscopy naturally depends on the morphology, state of differentiation, and function of the tissue. The lesion may be infiltrative or destructive. An example of the former occurring in the cornea is arcus senilis; depigmentation in the iris is a manifestation of the latter. The symmetrical aspects not only of normal features, such as the design of the pupillary pigment excrescences or details of iris color, but also the exact similarity in appearance of such changes, as senile atrophy of the pigment border, pingueculae, and cataracts in uniovular twins, as reported by Vogt, strongly indicates that so-called senile changes are genetically determined. On the other hand, it is possible that outside or accidental influences may modify, retard, or hasten the appearance of such changes. As Duke-Elder so aptly puts it "the processes of senescence should rather be calculated in terms of intensity of living, of heredity, and of the cumulative effects of rude shocks or quietly progressive toxemias."

Biomicroscopy affords us an excellent means of observing the "aging" processes in the iris which may involve all or any part of its mesodermally or ectodermally derived tissue. In a general way senile

changes in the iris resulting in atrophy of the stroma, degeneration, and depigmentation of ectodermal pigmented epithelium and the tendency of dispersal or even proliferation of pigment may be classified according to the parts chiefly affected as: (1) changes of the pupillary pigment border (seam); (2) circummarginal border changes adjacent to the seam; (3) rarefactions of stroma and posterior retinal pigment layers distant to the pupillary margin.

Senile Changes of the Pupillary Pigment Border. The principal and most typical senile change in the iris pigment seam is one in which destruction and migration of pigment occurs (Plate XLIII, fig. 1).

Because the pupil tends to become smaller with age and with adjacent stromal changes, it is not uncommon to see an increase in prominence of the pigment excrescences of the pupil border during middle life and even occasionally in the aged. As a rule, however, with the passage of time, owing to disintegration and dispersal of pigment a segmentation of the seam occurs which is followed by a progressive decrease in the number of excrescences so that in certain areas they may completely disappear. Such changes may be minimal or may progress until the seam is completely destroyed. The pigment itself, being free, may be transported by the aid of the aqueous and gravity and be deposited on the posterior corneal surface, the anterior lens capsule, the surface of the iris, in the chamber angle, and even on the remaining pupillary excrescences, forming nodes. The deposition of pigment particles on the excrescences may cause them to have a dentate appearance. Pigment may migrate superficially within the stroma itself or may be deposited on the surface among the trabeculae in various guises, e.g., spheres, clumps, or particles (Plates XLIII, fig. 2; XLIV, fig. 3). An apparent disappearance of the pigment seam, simulating senile destruction, may occur with posterior synechiae following iritis. If the adhesion of the iris pigment border to the anterior lens capsule occurred while the pupil was partially dilated, it might become inverted or rolled in when the iris regains its mobility and the pupil becomes smaller. However, dilatation with a mydriatic would reveal the presence of the seam

at the point of attachment to the capsule. Such an inversion of the pupillary border may include part of the sphincter at the site of the synechia and cause the sphincter to be narrowed or absent. But,



FIG 315

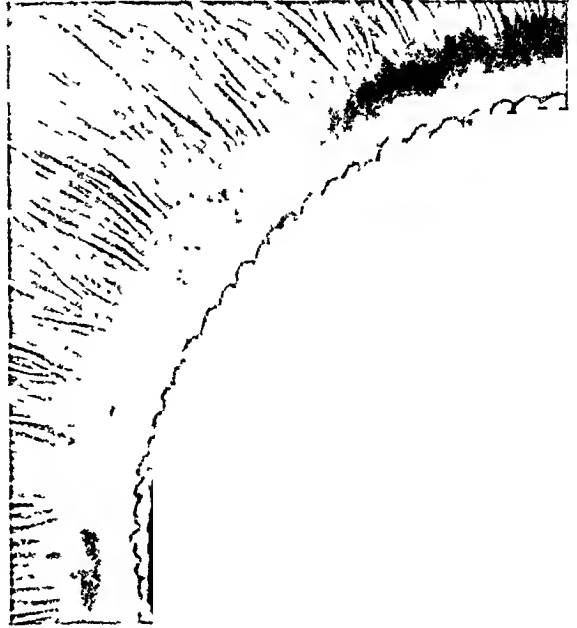


FIG. 316

FIG. 315. Senile atrophy of pupillary border and stroma as seen by retro-illumination. Note lacunae resulting from loss of substance; also migratory secondary pigmentation.

FIG. 316. Senile circumferential sulcus. (After Vogt.)

again, dilatation of the pupil reveals the actual condition (posterior synechiae) and the sphincter edge reappears or widens.

Senile atrophy may result in a rarefaction of the pupillary margin which is easily seen by means of retro-illumination (Fig. 315). To demonstrate the atrophic nasal iris border by this method the light beam is diverted from the temporal side into the pupil so that it passes obliquely into the lens in the vicinity of the nasal border. Conversely, to examine the temporal side of the iris the process is reversed so that the beam is directed temporally from the nasal side. If the pupillary marginal pigment is intact no retro-illumination occurs. However, if rarefaction of the retinal pigment has occurred the border may appear moth-eaten or delicate translucent pearly-gray or blue areas may be seen like empty filmy envelopes. The normal contour of the pupillary margin is retained, appearing in the

later stages of this process as a whitish veil somewhat glassy in texture.

Occasionally, a senile hypertrophy of the pupillary pigment seam may be seen instead of atrophy (Plate XLIII, fig. 4). This change, which is more likely to occur above than below, is characterized by a sharp angulation between the enlarged individual pigmentary excrescences. Such a condition may not be due entirely to an overgrowth but rather to a slight eversion as occurs in miosis (since the pupillary diameter is always narrower in senility) augmented by traction exerted by sclerosed stromal fibers which are unable to stretch or elongate sufficiently as the pupil gets smaller.

Senile Circummarginal Changes (Adjacent to Seam). Gutter formation (sulcus circummarginalis of Vogt), atrophy and destruction of the stroma and posterior retinal pigment layer can occur, immediately adjacent to the pupillary seam (Plate XLII, fig. 2). The area involved is rarely more than from 0.1 to 0.5 mm. in width (Vogt) and is best seen in light-colored irides. At times a narrow, glassy-looking furrow, partly or completely encircling the pupillary border, may be seen (Fig. 316). The walls and floor of this gutter have a homogeneous character. It is usually impossible to see any of the normal radiating stromal trabeculae within it. In some cases it seems as if the radiating fibers stop at its distal edge. In blue irides this furrow appears yellowish in color and in brown irides it is less pigmented than the surrounding stroma. The pigment excrescences may be missing or only slightly attenuated. Frequently, I have seen such circummarginal changes without any depression or gutter. This may represent an interim stage of sulcus circummarginalis.

Concentric with the seam, there may be a narrow area of stromal atrophy through which the deeper and darker retinal pigment layer may be seen. Occasionally, a few strands of stromal fibers may traverse this atrophic area to the pupillary pigment seam. Senile atrophy and destruction of the pigment border and the adjacent posterior pigment layer may result in a marked rarefaction; this occurs in the form of irregular areas or triangular (cuneiform) sectors, the apices of which are directed toward the periphery, or in

the form of clefts, appearing almost as if caused by a lateral separation of the excrescences (Fig. 315). Such areas of destruction may be confined to isolated portions or may extend circumferentially around the border in a continuous or segmented manner. In some cases these marginal defects may extend peripherally in the posterior retinal layers as far as the region of the contraction furrows, and they may leave depigmented translucent areas in the form of radial streaks or irregular perforations which can be outlined by retro-illumination. The seam is usually missing in these areas and a fine dispersal of the pigment may occur in the neighboring stroma. The finding of such pigment dispersal in the stroma (particularly in blue irides) as a senile change tends to refute the idea of Koeppe that by itself it represents a preglaucomatous change. In ordinary direct illumination or daylight, as a result of reflection from residual uncolored stroma, it may seem that except for an absence of excrescences the border is normal. However, retro-illumination, especially if aided by a cataractous lens, discloses that the iris margin of the affected areas consists of nothing more than a diaphanous veil, devoid of retinal pigment and with only a film of stroma remaining. By daylight or ordinary oblique illumination the iris border may appear somewhat whitish and hence before the days of biomicroscopy (retro-illumination of the iris border by lens light), it was incorrectly interpreted as being due to a "hyalinization" of the pupillary margin.* This type of atrophy of the pupillary margin is similar to that which occurs after chronic iritis and especially in heterochromia iridis.

Senile Rarefaction of the Stroma and Layer of Retinal Pigment Distant to the Pupillary Margin. With age, especially in those lightly pigmented cases in which the stroma trabeculae can be seen, one

* It is extremely doubtful whether the so-called hyaline degeneration of the pupillary margin of Fuchs (1884)⁴³⁶ can be seen biomicroscopically. Pathologically, it consists of a hyalinization of the stroma and adventitia of the vessels, especially behind the sphincter and is supposed to result in senile miosis and rigidity of the pupil. In all simple senile cases which I examined, in which there was a whitish band at the pupillary margin, retro-illumination revealed destruction of the pigment seam with the resulting diaphanous veil. In several cases the whitish tissue consisted of flakes of exfoliated lens capsule (seam intact), though in two of these (with normal pressure) no areas of exfoliation could be seen in the central exposed parts of the anterior lens capsule.

becomes aware of definite thinning of its entire structure, although the individual trabecula may become more fibrous and less transparent, resulting in a loss of brilliance of the entire iris surface. For example, a blue iris tends to become duller and more gray in the aged.

Occasionally in light blue irides of the aged one can see a dark band between the distal edge of the sphincter and the frill, anywhere up to 0.5 mm. in width. This is probably caused by atrophy of the stroma which permits the dark unaffected posterior layers to shine through.

Another frequent type of senile atrophy or degeneration of the mesodermal tissue, also seen in blue irides, is a form in which there is rarefaction of the deep stroma, immediately over the sphincter itself (Plate XLIII, fig. 3). This causes the sphincter area to appear brownish in color. This brown area may be crossed by a few residual fibers, extending from the frill to the seam, but in any case these fibers are not sufficient in number to prevent the deep brown color of the posterior layers from coming through.

In extreme age, especially in blue or gray irides and when the pupil is small, one gets the impression that the radiating trabeculae are less wavy and more attenuated and that the supporting tissue is more opaque and collagenous. This form of senile "sclerosis" causes a flattening of the iris surface with a tendency toward obliteration of the surface markings and crypts. At times this condensation of the iris stroma results in a loss of demarcation between the pupillary and ciliary zones (lesser circle), the sharp slope of the pupillary zone toward the pupil no longer being noticeable. The irregular (zigzag) line of the frill itself may become so attenuated into the hyaline stroma that it is seen with difficulty. However, this type of sclerosis (in light colored irides) never obscures the sphincter but rather makes it more visible owing to the thinness of the overlying stroma. Such atrophic changes may involve the entire iris uniformly and may cause a change in its color depending on the amount of pigment destruction. An increase, however, of stromal atrophy in localized areas may develop to the point where the posterior pigment layers

are exposed to view. At times only a few atrophic trabeculae may be found crossing the exposed pigment layers. Rarely, their curled detached ends may float freely in the chamber, giving the impression of a localized detachment of the stromal fibers.



FIG. 317. Senile rarefaction of posterior pigment layer as viewed by retro-illumination. The cataractous lens acts as a strong reflecting surface.

The more extensive senile tears or separations, especially of the superficial stromal leaf, have been described by Schmitt, Mlle. Drapkin, Sander, Dollfus,³⁰⁹ and Vogt. Such separations indicate the morphologic and genetic independence of the superficial stromal leaf and remind one of other more or less spontaneous detachments or separations, e.g., of the lens, retina, vitreous, all surrounded by fluid having a lower specific gravity.*

Vogt has pointed out that occasionally in the pupillary area or even more so peripherally, especially in the crypts, small black points may be seen, particularly in poorly pigmented irides. These small black points represent rarefied areas in the posterior pigment layers.

* Vogt believes that there is a hereditary disposition to atrophy or cyst formation and that gravity together with "eye movements" as a *vis a tergo* are sufficient to cause these progressive separations. He has divided these separations of the iris stroma into two types: first, in which a separation of the superficial layer of stroma occurs, and second, in which there is a separation of the entire stroma (superficial and deep). When the separation is marked the stromal fibers may be seen floating freely in the anterior chamber. Some may even be in contact with the posterior corneal surface. Separation of the mesodermal layers almost always exposes the dark brown posterior pigment layers to view.

Larger areas of rarefaction of the posterior pigmented iris layers may be seen with retro-illumination in the form of round holes, radial streaks, or as an irregular circumferential band in the extreme iris periphery near the limbus.* This retroreflected light, passing through unobstructed, outlines these defects, causing them to stand out lighter in color than the darker adjacent unaffected areas (Fig. 317). Atrophic areas in general are better seen when the lens is cataractous because the opaque lens reflects more light than a normal one (Fig. 317). The frequency of iris atrophy, associated with ordinary senile opacifications of the lens, comparable in a way to the findings in the syndrome of heterochromic iridis (Fuchs), is well known.

Senile Alterations of the Iris Sphincter. Changes of the iris sphincter due to age are best seen in blue or gray irides, especially when associated with rarefaction of the stroma. The sphincter appears to have an increased density which often can be seen as well by direct focal illumination as by indirect illumination. Depending on the nature of the surrounding tissue its color varies from a dirty gray to yellow or even pinkish. Frequently the sphincter appears to be narrower in age.† This may be sectorial or circumferential. Koby⁵⁰¹ states that "in marked cases, it gives the impression that the sphincter has shrunk away from the peripheral part of its bed, which then becomes visible as a darker ring." Local narrowing of the sphincter, its width being reduced to half, may occur in the region of an extensive posterior synechia following long-standing iridocyclitis. Occasionally in a blue or gray iris of the aged a dirty gray cleft separates the pigment border from the sphincter, resulting in a sphincter-free annular zone somewhat similar to that observed in circummarginal destruction of the pigment border. Vogt has described senile atrophy of the stroma in a case of compensated glaucoma in which there were narrow stripes of pigment within the sphincter.

* It is not surprising that the periphery of the iris manifests rarefaction since the posterior pigment layer is thinnest there.

† On the other hand, in buphthalmos widening of the sphincter up to 2 mm. has been noted.

Chapter Nineteen

CONGENITAL ANOMALIES OF THE IRIS

THE fact that embryologically there is considerable difference in time of appearance and development of the mesodermal and ectodermal iris layers * has resulted in many conflicting explanations concerning the evolution of iris anomalies.

In discussing developmental defects of the iris, Ida Mann states that "with regard to the interaction of the mesodermal and ectodermal portions of the iris in normal development not much is known. It is generally accepted that the mesodermal portion, being formed first, acts as a sort of scaffold or directional tissue plane for the subsequent forward growth of the pigment epithelium which lines it later. On the other hand the optic outgrowth can be experimentally isolated *in vitro* and will invaginate and form a retina and iris (ectodermal only and not usually completely formed) without contact with mesoderm at all. It is probable that both tissues are self-determining though the possibility of chemical interaction between them must be admitted."

Recently, in a family of 8 it was found that 4 members had some form of persistent pupillary membrane. Many writers in this field have held that hereditary transmission of these defects is direct. On the other hand, considering the existing evidence one cannot entirely deny the influence of such factors as intrauterine inflammation; various mechanical factors and metabolic interferences (chemical, toxic, nutritional,† or glandular dyscrasias). The importance of the endocrine secretions in the production of many types of anomalies

* The ectodermal (posterior layer) projection occurs at about the 65 mm. stage, at a time when the mesodermal framework is well determined.

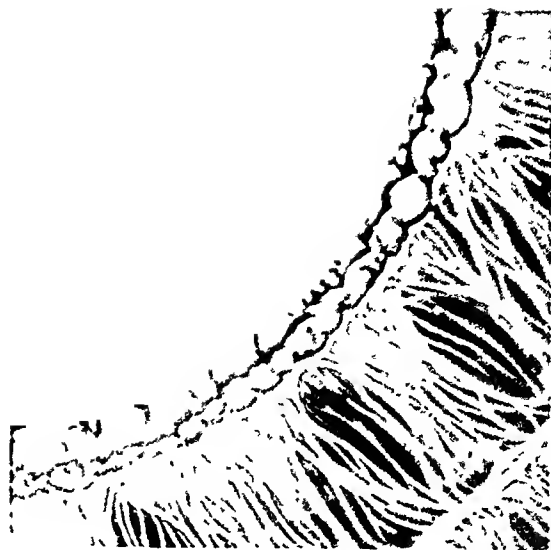
† Warkany, (Vitamins and Hormones, Vol. III, 1945), produced congenital defects of the eye by means of nutritional deficiencies in the mother.

PLATE XLIV

- FIG. 1. Hypertrophy of the pigment seam of the pupillary border.
- FIG. 2. Double seam.
- FIG. 3. Particle of detached pigment of the seam resting on the iris surface.
- FIG. 4. Tuberous flocculus.
- FIG. 5. Pendulous and exfoliative flocculus.
- FIG. 6. Pilocarpine cyst of the pupillary pigment.
- FIG. 7. Cystic degeneration of the seam in diabetes.



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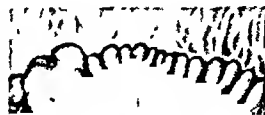
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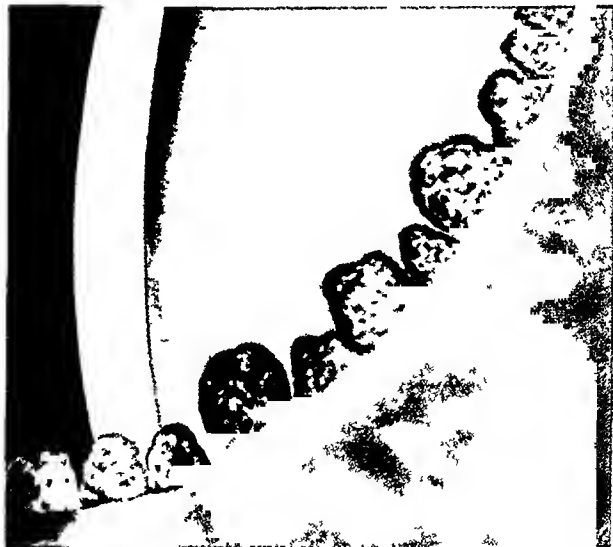
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is uncertain, considering the fact that many of these glands do not actively function until after the development of a specific type of tissue has occurred. With regard to the influence of aberrant bands or persistent vessels (persistent capsulopupillary vessel), there is clinical evidence that in occasional cases a causal relationship is indicated by their presence.

Because it is not always possible from clinical appearances alone to explain which of these factors are responsible for the production of the different anomalies of the iris, one can only formulate a classification of these defects on the basis of extent of involvement of, or selectivity of the affection for, one or both germinal layers. Such a classification must inevitably be faulty because it does not take into consideration possible relationship between incompletely developed ("*formes fruste*") and completely developed forms. For example, if iris defects are divided into two groups, (1) those involving its whole thickness and (2) those involving one type of tissue predominantly (Mann), it would follow that an incomplete coloboma (defect of the mesoderm only) could not be placed under the same heading as a complete one (localized absence of both layers) even though the same etiologic factors were at work in both cases. Consequently in descriptions of these anomalies a strict classification will not be adhered to, and only those defects that are common or interesting from a biomicroscopic viewpoint will be dealt with.* Since the advent of biomicroscopy many minute "subclinical" anomalies of the iris have been brought to our attention.

ANOMALIES OF THE PUPILLARY PIGMENT BORDER (SEAM)

Because the seam is so commonly less well developed inferiorly than superiorly, such an inequality may well be considered as falling within normal limits. The same holds true for the occasional absence of an individual pigment excrescence at the lower border. However, a majority of the malformations of the pigment border are due to localized hypoplasias of the retinal pigment layer.

* For those desiring a more exhaustive handling of these anomalies I suggest perusal of special treatises on this subject, and especially recommend to English readers Ida Mann's "Developmental Anomalies of the Eye."⁶³⁵

Congenital reduplication (doubling) of pigment border is an anomaly which according to Vogt consists of a doubling of the pigment border, is not rare and may be hereditary (Plate XLIV, fig. 2). An interrupted circular furrow seems to divide the seam into an axial and peripheral part. The width of the reduplicated seam is usually not more than 0.1 to 0.2 mm. Reduplication of the seam may be complete or incomplete. In the former, it extends circumferentially around the border, while in the latter it may be localized to a sector as short as 1 mm. in length. This anomaly is probably due to an extension of the folds of the retinal pigment layer which, on reaching the pupillary border, evert and become visible as an extra seam.

Flocculus iridis is characterized by a hyperplasia of the retinal pigment at the pupillary border, appearing as nodular "grapelike" clusters or as if the hypertrophied seam were twisted on itself like a hempen rope, or as flattened and twisted elongated pendulous processes (Plate XLIV, fig. 4, 5). It is generally conceded that flocculus formation is definitely hereditary (but not sex-linked) in character. This condition must be differentiated from ectropion of the retinal pigment in which the everted retinal layers extend over and lie flatly attached to the mesodermal iris layers. Although in both these anomalies (similar to the normal seam) pigment excrescences appear black in daylight, the focal beam reveals them as dark brown in color. The fact that in man flocculi are generally situated above has caused them to be likened to the pigment processes normally hanging over the pupils of horses and cattle and to be considered atavistic vestiges. However, anatomically, flocculi in man do not have the core or substratum of connective tissue found in animals; Fuchs⁴³⁴ has shown that they consist merely of a hyperplasia of the pigment epithelium with no enclosed connective tissue. However, the tuberous type in man may have connective tissue strands (iris stromal fibers) connected to them. Ida Mann has suggested that this may represent a similarity to flocculi seen in animals, in which these structures become invaded and supported by mesoderm. It is possible that the form examined histologically by Fuchs was not of the tuberous variety.

Based on clinical appearances, Vogt and others consider that there are two distinct types of flocculus iridis; both may be found together: (1) Tuberous (*flocculi tuberosi*) and (2) exfoliative and pendulous (*flocculi exfoliativa et pendulantes*). The first type, com-



FIG. 318. Tuberous flocculi at upper border of the pupil.

posed of irregular humplike or globular pigment masses, usually has attached (mesodermal) stromal fibers (Fig. 318). Frequently one sees fine nets of trabecular iris tissue over the flocculi which extend peripherally to the frill. In others the frill may be absent or may be drawn up over and attached to the convoluted pigmented mass. In all cases, it should be remembered that the condition, although involving the seam, actually develops from the posterior retinal iris layer. It may be, as suggested by Vogt, that owing to an abnormally strong fixation between the frill and the retinal iris layer in the fetus, the deeper stroma fails to extend axially to a sufficient degree, causing traction with subsequent eversion and overdevelopment of the underlying pigment layer. Koby states that this form "owes its origin to a nipping off of a mass of pigment by fibrous processes of the mesoderm."

The second or exfoliative and pendulous type is free of stromal attachments, and the flocculi appear as "flattened convoluted lamellae" of smooth surface or, owing to cystic degeneration, they form large globules resembling grape seeds. Several of these dark

brown rounded globes may fill the pupil. Some may be attached to the seam and others may take origin from the posterior surface of the iris. Although both forms may remain stationary throughout life, cystic degeneration in the second form may ensue with enlargement,



FIG. 319. Pendulous form of exfoliative flocculi. (After Vogt.)

possibly leading to the pendulous kind, since frequently both (exfoliative and pendulous) are seen in the same iris.

A variant of the exfoliative type is the extreme pendulous form in which the masses may be extended in the shape of Indian clubs (Fig. 319). These elongated processes resemble the flocculi seen in animals. Unlike the tuberous variety, they are almost always found superiorly. There they are not derived from the pigment seam (which may be normal at the place where the process is attached) but rather from the posterior retinal layer behind and above it, thus hanging suspended between the lens and iris. Cases have been reported in which pendulous flocculi were held suspended by a necklike attenuated thread. This may possibly be a transition stage in their development, ending by detachment of the head, which in turn may be found in the chamber angle as a solid pigment globule or more often

lying loosely adherent to the surface of the iris. In a few cases they have been seen floating freely in the anterior chambers.* It is not unlikely that a portion of a flocculus itself may break off directly without transitional elongation. In one instance it was noticed that such a corpus mobili disappeared from view (broken down and eliminated ?). However, globules of pigment of similar appearance, lying on the iris stroma, have been seen in the absence of any flocculi formations. These detached pigment bodies may assume shapes other than spherical, appearing, for instance, flattened or bean-shaped, and though they are generally of a solid consistency, cystic degeneration may cause them to be translucent. It has also been suggested that cystic flocculi (free or attached) may be associated with a persistence of the marginal sinus.†

Pilocarpine Cysts of the Pigment Border. The prolonged miotic action of pilocarpine or eserine (physostigmine) may result in the formation of cysts of the retinal pigment layer (Vogt). These are seen as pigmented globules at the pupillary border of the iris. At times these structures resemble small pouchlike bags of pigment suspended from the puckered pigmented excrescences (Plate XLIV, fig. 6). These cysts may arise from the posterior surface of the iris just behind the pupillary seam so that their attachments are hidden, or they may arise from the excrescences themselves. In the former case the pigment excrescences may be normal, and the cyst may displace the iris border away from the lens. At the base or place of attachment small furrows may be distinguished but the ballooned-out portion is generally smooth. In some, the surface is smoothly pigmented, while in others it appears granular as if dusted with pigment. In some cases only a single cyst may be found, while in others the entire circumference of the pupillary margin may be dotted with them.

* Cases have been reported in which these bodies were attached to remnants of the pupillary membrane and consequently were considered as being derived from mesodermal tissue. But it is also possible that a detached ectodermally-derived pigment globule may have become adherent to a pupillary membrane. Histologic evidence in almost every case has shown that these bodies are composed of cells resembling the posterior retinal epithelium of the iris.

† The marginal sinus is a circular space between the two layers of the rim of the optic cup. It remains open until the end of fetal life, ultimately closing by fusion of its walls.

It is interesting to note that this type of cyst is invariably related to pilocarpine or eserine miosis and is not found in untreated cases of glaucoma. The discontinuance of the use of these drugs may cause them to disappear. At times the cysts may vanish through bursting and in some instances they reform later. A shrunken or burst cyst may resemble a flocculus. As a rule, they can be differentiated from flocculi by the fact that cysts are distended by fluid and can be transilluminated if they are not too densely pigmented. Also, cysts are attached by a narrow contracted base and have ballooned-out bodies. Especially important is the fact that these cysts may be transient. The tendency of the retinal pigment to form cysts suggests that this tissue may have a secretory activity. This activity may account for the formation of the larger congenital cysts associated with the pigment (neural) epithelium on the posterior surface of the iris. The latter may be due to a persistence of the annular sinus or to the migration of clumps of retinal epithelium into the mesodermal stroma with secondary cystic degeneration.

Diabetic Changes in the Pigment Excrescences. Histologically, it has been known for a long time that swelling, cystic degeneration, and detachment of the pigment-bearing epithelium of the posterior iris surface and of the pupillary excrescences may occur in diabetes (Fig. 320). Likewise, when performing an iridectomy in diabetic patients it has been noted that manipulations of the iris may cause a bursting of these cells with the resultant formation of a black or dark brown cloudiness in the anterior chamber owing to dispersal of the pigment.* With the biomicroscope, evidence of such edematous or degenerative changes in the retinal iris pigment will often but not invariably be seen in diabetic patients. When the pigment excrescences of the pupillary border are so affected they are enlarged and have a spongelike or porous aspect, appearing pitted (Plate XLIV, fig. 7). This rarefaction can frequently be demonstrated by

* The sudden appearance of a dark cloud (evidently fucsin) in the aqueous during mydriasis in cases of senile exfoliation of the anterior lens capsule has been seen by Vogt in nondiabetics. These cases were complicated by glaucoma, and the patients had been subjected to prolonged miosis. In these instances the cloud might result from the bursting of (pilocarpine) pigment cysts, although in a case of untreated chronic simple glaucoma a similar occurrence took place.

retro-illumination, in which case the pigmented border may appear to have a lacy texture. At the site of a posterior synechia the attached porous excrescences may be drawn out into dentate pyramidal processes. In some instances the pigment border itself may be unaffected but retro-illumination may reveal atrophy of the posterior pigment lining in the form of radial or irregular lacunae distal to the pupillary margin; these lacunae resemble senile or post inflammatory atrophy of the iris. As a rule little change is noted in the stroma but free pigment granules may be scattered over its surface.



FIG. 320. Histologic preparation of cystic degeneration (glycogen infiltration) of the pigment-bearing epithelium of the posterior iris surface (diabetes). (After Gilbert.)

*Ectropion (Hyperplasia) of the Retinal Pigmented Layer of the Iris.** Although not uncommon in pathologic states of the iris, e.g., absolute glaucoma (page 868), ectropion of the retinal pigment layer rarely occurs as a congenital malformation, in many instances being of proved hereditary origin. The presence of this anomaly in identical twins (Vogt) seems to indicate beyond controversy that this is true. However, it is probable that fetal inflammation may

* The older appellation, congenital ectropion of the uvea, should be discarded since the everted pigment is ectodermal in origin.

PLATE XLV*

FIG. 1. Congenital ectropion of the pupillary border with distortion of the pupillary outline.

FIG. 2. Ectropion of the pigment of the pupillary border of the iris.

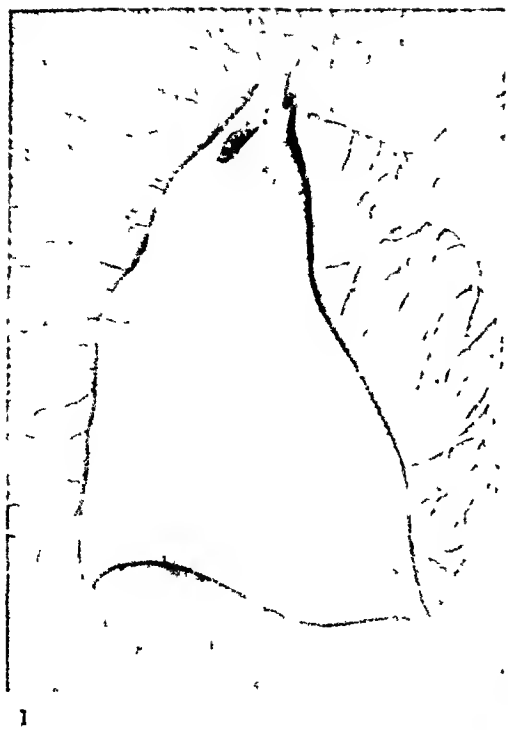
FIG. 3. Ectropion of the pigment border of the iris (apron form) with area of melanosis iridis.

FIG. 4. Partial coloboma with pigment stars on the anterior lens capsule.

FIG. 5. Aniridia. Observe the zonular fibers and the unusual type of lamella cataract. Right eye.

FIG. 6. Same case as shown in Figure 5. Left eye.

* Due to editorial changes this plate was originally Plate V (see opposite page).



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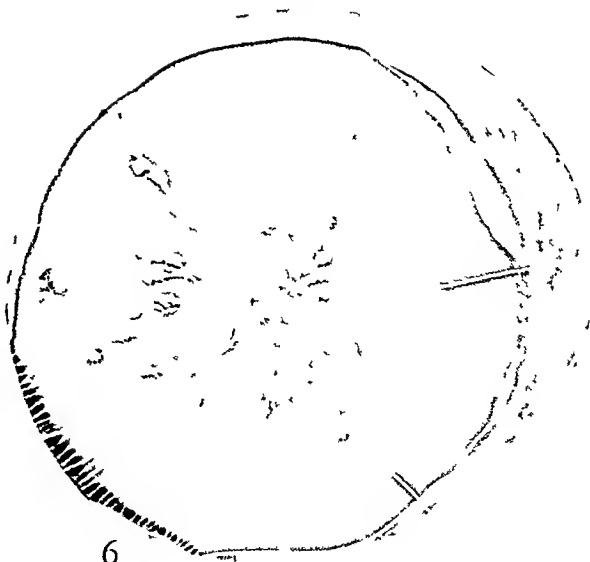
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be a causal factor in isolated "congenital" cases. Ectropion of the retinal pigmented layer of the iris is similar to flocculus in that there is an extension of the border pigment; it differs from flocculus in the fact that in ectropion the pigment never hangs free in the anterior chamber but lies flatly attached to the iris stroma. Moreover, ectropion has no predilection for the upper pupillary border but may occur anywhere on the iris.

Two forms of this anomaly may be seen: (1) ring form in which the pigment eversion (generally narrow in width) extends completely around the pupil; and (2) apron form in which a partial sectorial overlapping of the stroma occurs. On hasty examination students have been seen to mistake the first for an enlarged pupil and the second (when large) for a coloboma (pseudocoloboma),⁵⁰² or even an irregular pupil (iritis). Such errors naturally would occur only when a superficial examination was made by daylight and would be impossible with biomicroscopic inspection.

The degree of ectropion varies from case to case (Plate XLV, fig. 1, 2, 3). In some, all that is seen is a small eversion from 2 to 3 mm. in extent, at any point of the marginal circumference. Such small eversions may not reach the frill and their surface is pigmented corresponding to the serrations of the seam. As a rule, the eversion seldom extends beyond the frill (although cases have been noted in which the pigment has extended over the sector of the iris as far as the periphery). The smaller ones, 0.5 to 1 mm., may disappear during mydriasis. The surface of the ectropion is usually smooth in the area away from the rounded and slightly dentate pupillary margin, though on occasion radiating serrations may extend peripherally for a considerable distance (Fig. 321).^{*} All these serrations or folds tend to flatten out with mydriasis. At times I have noted small lump-like pigmented surface nodules in distal parts. The fact that there is always perfect action of the pupil in the involved area seems to indicate that the sphincter is not involved in this condition. Occasionally one will notice a small thickened area of hyalinization of the

* Pathologic ectropion is characterized by little or no striations at the pupillary margin, as well as by enlargement of pupil and extensive stromal changes.

iris stroma near the distal apex of a slight ectropion, just in front of the frill, resembling a scar. In some only a few pigment granules may be seen in the stroma scattered in the vicinity of the eversion;

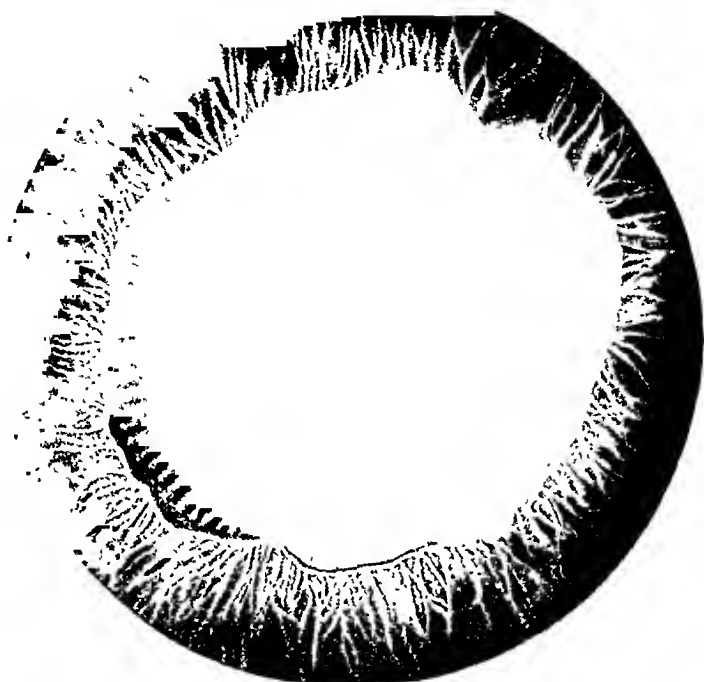


FIG. 321. Ectropion of retinal pigment showing radial serrations at the pupillary margin.

in others a considerable area of lighter brown pigmentation is found surrounding the ectropion.

The exact mechanism involved in the production of this anomaly is imperfectly understood. Normally the double layer of pigmented epithelium follows the mesodermal scaffolding until it reaches the pupillary margin and forms the pigmented seam as seen from in front. It is conceivable that if the growth of stroma in the pupillary area is retarded, the extension of the posterior pigment layer can advance a little ahead of it and can extend around the anterior surface until it meets the delayed superficial stromal growth. Moreover, it has occurred to me that if the terminal end of the retinal pigment layer (marginal sinus) became attached to the back of the embryonic pupillary membrane, it might be drawn back by traction when atrophy and retraction of this membrane occur during further development. An adhesion of this type in association with a cystic

detachment of the marginal sinus might also be a factor in the production of flocculus.

DEFECTS PRINCIPALLY INVOLVING THE STROMA

Minimal Defects of the Stroma. Minute defects of the stroma are seen occasionally. They may be localized or appear in several areas. One example of the latter is found in cases in which fine stromal trabeculae extend over the pigment border in the form of hooked strands very much like clasps. Still another is that in which the frill (lesser circle) is in contact with the seam itself (Plate XLI, fig. 9). Anterior adhesions of the iris stroma to the posterior corneal surface may occur in the periphery, or extensions of the frill may be attached to the pupillary area of the posterior corneal surface.* Vogt has described small nodes of condensed tissue lying on the iris surface at times associated with a gap in the seam and ectropion. Other local condensations resemble scars and exert traction on the frill (Plate LVI, fig. 2). In these defects the surface tissue appears clouded so that the radial strands are indistinct. Occasionally a vessel may be present. In another case described by Vogt there was a small atrophic area of stroma with exposure of the posterior pigment layer. At this point there was an absence of the sphincter tissue, indicated by a lack of pupillary action. The rarefaction was in the horizontal meridian so that it bore no relation to the fetal cleft. Such local mesodermal thickenings or atrophies suggest inflammatory lesions during fetal life (fetal iritis).

Occasionally in blue irides an increase in stromal pigmentation in the region of the frill may be observed. This tends in rare cases, according to Vogt, to form a light yellow or orange colored ring regularly concentric with the pupillary margin. In other instances, more common than the above, a heavy, irregular or spotty brownish pigmentation may occur in the superficial leaf of a blue iris near the frill. These pigmented or melanomatous spots resemble pigmented nevi. When such spots are found in the pupillary zone (deep stroma) they may be derived from the clump cells of Koganei, normally seen

* See Volume I, Plate XXXVIII, fig. 3.

in histologic preparations near the sphincter. It is not unusual to see darkened clumps of pigmented cells in the superficial stroma of brown irides, but when such spots are scattered over a light colored iris (blue or gray), the iris has a greenish hue (piebald iris).

Physiologic Rarefaction of Stroma. If one studies the deep blue irides in daylight, a dark sector or shallow groove will frequently be seen in the ciliary zone directly at 6 o'clock (Plate XLI, figs. 3, 4). This is caused by a hypoplasia of the stroma in the region of the fetal cleft and may be considered as a physiologic fetal rest or possibly as a rudimentary coloboma. In Italians, Streiff found it in 25 per cent of normal irides examined. This defect is best noticed macroscopically by diffuse illumination or even better by daylight, and may appear as a large crypt or as a sector of rarefied stroma which permits the deeper retinal layers to show through.* In the region of the defect the frill and contraction furrows may be absent. In one case examined by the author there were no pigment excrescences at the pupillary margin of the affected part. The close relation between physiologic rarefaction and incomplete colobomas is well illustrated in a case reported by Mann. In this case there was a physiologic rarefaction of the stroma below in one eye, while in the other a similarly situated rarefaction was accompanied by a small notch of the pupillary margin. In addition, retro-illumination produced transillumination of the involved segment, indicating partial absence of the retinal pigment layer.

Coloboma Iridis. Coloboma iridis is a common developmental anomaly of the iris and is usually hereditary, although it may occasionally result from other causes. In contrast to aniridia (page 799) in which there is an almost complete absence of the iris, the term "coloboma" signifies the absence of one or more sectors of the iris. The literature describing iris coloboma is replete with classifica-

* This type of physiologic rarefaction of the stroma, however, differs from a Fuchs' crypt. Fuchs' crypts are found in any meridian of the iris surface, and only involve the superficial stromal leaf. Also their margins are sharply outlined by the trabeculae of the superficial leaf and they rarely extend to the ciliary iris margin. Physiologic rarefaction, on the other hand, only occurs below in the region of the cleft, extends from the seam to the ciliary margin, and involves the stromal thickness, its boundaries subtly merging with the unaffected neighboring iris tissue.

tions and subdivisions depending on the location, size, or tissues involved. For instance, a coloboma is regarded as being typical when it is located below in the direction of the fetal fissure or cleft. This

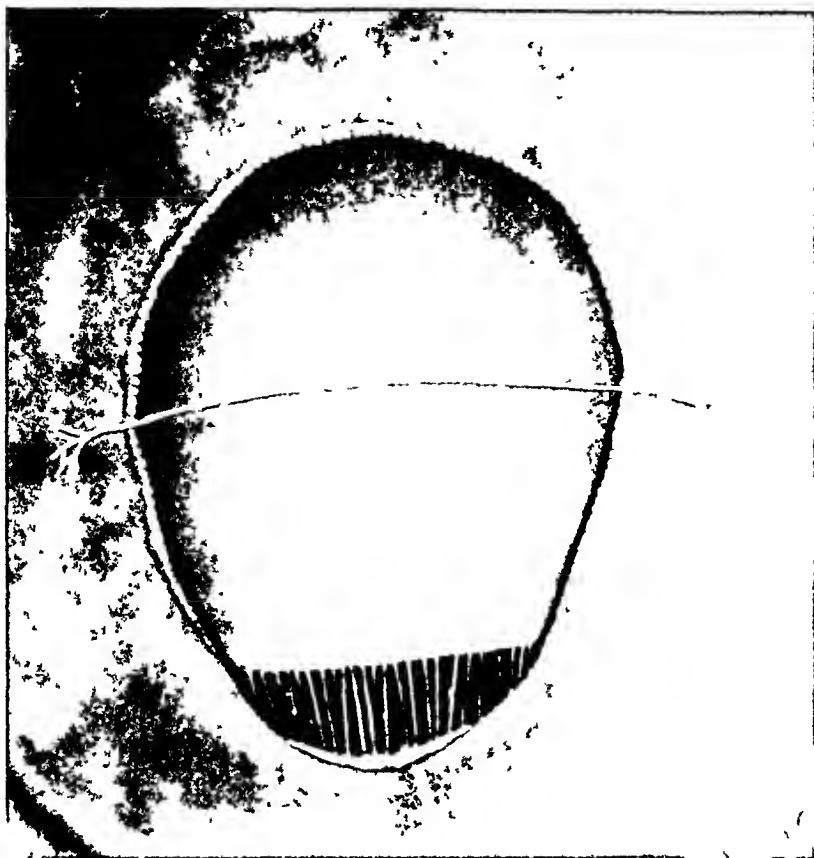


FIG. 322. Incomplete coloboma (partial) with rests of the pupillary membrane.

form, which commonly coexists with a similar defect in the ciliary body and choroid, is considered as an iridic extension of these defects. Located in any other place a coloboma is called "atypical." When a coloboma involves the whole visible width of the iris, it is considered total, otherwise it is partial, e.g., a notch of the pupillary margin, a hole (false polycoria or dehiscence—congenital iridodialysis or diastasis). If the defect affects both mesodermal and ectodermal layers or the entire iris thickness, it is complete, but if only one or the other is missing it is an incomplete coloboma or a pseudocoloboma (see physiologic rarefaction of stroma) (Plates XLV, fig. 4; XLVI, fig. 6).

In most cases, whether total or partial, colobomas are triangular or pyriform in shape, the apex pointing toward the ciliary border (Fig. 322). Typical colobomas usually tend to be total but their pillars may be spanned by mesodermal strands or bridges. Such a bridge at the ciliary border below may separate the iris defect from that in the ciliary body. The exact appearance of the pupillary borders differs from case to case. As a rule there is a continuation of the seam from the unaffected parts of the iris but the excrescences may be thinned out or flattened in some areas or missing in others. It is not unusual to find the pigment beading defective or missing on one pillar only. In some there may be an irregular hyperplasia of the seam, not unlike an ectropion. Although the stroma is usually normal adjacent to the seam, on one side or the other a narrow whitish line of atrophic tissue may be seen. The frill usually stops sharply at the margins of the coloboma as if it had been cut off; in cases in which there is only a notching (partial coloboma), the frill may continue around the defect and be attached to the seam. The sphincter is usually absent in the region of the defect. Commonly there are remnants of the pupillary membrane attached to the stroma at the sides of the "legs" of the coloboma. Persistent pupillary membranes may be associated with or connected to starlike pigment deposits on the anterior lens capsule or such pigment deposits may occur without the presence of membranes. The edge of the lens (equator), when seen, may appear crenated or wavy, especially when gaps or defects of the zonule are present. Zonular or tenuous lamellar opacities frequently curve around the exposed equatorial parts of the adult nucleus of the lens. In a case recently seen with similarly situated opaque lens fibers, there was a pyramidal anterior polar cataract.

The pathogenesis of colobomas of the iris is not completely understood but there are two chief possibilities: first, that the defect is primarily ectodermal owing to a local failure in development of a sector of the optic cup margin; and second, that it is primarily mesodermal, owing to the inhibitory effect of persistent capsulo-pupillary vessels. Mann states: "A number of arguments can be found

on both sides and it is probable that both types of defect occur, the assigning of a cause in a given case to either group depending on minute details of individual structure."

ANIRIDIA CONGENITA

Generally the term "aniridia" would suggest a complete absence of iris, but actually, as indicated by all of the cases of this irregularly dominant hereditary anomaly reported in the literature, some rudiments or stumps of iris tissue were found. Usually a narrow rim of poorly developed iris, either sectorial, or partially or completely annular, forms a pupillary margin. This rim, although markedly underdeveloped, contains both mesodermal and ectodermal elements. However, no sphincter or deeper stromal layer is found. Mann states: "The absence of the deeper portion of the stroma is easily understood since this develops later in response to the need of the sphincter for a blood supply and is presumably initiated by the differentiating rim of the cup. If this failed, the sphincteric and subsphincteric plexuses and thin layer of feeding vessels would be absent too."

Biomicroscopically the rim or iris stump can easily be seen unless it is entirely hidden behind the corneoscleral margin (Plate XLV, figs. 5, 6). However, in most cases if the direction of the beam is widely angulated and the patient's eye abducted or adducted, some vestige of the stump can always be seen. Its presence can also be demonstrated by gonioscopy. Nystagmus often makes examination difficult. The pupillary margin of the iris stump may show normal pupillary excrescences, part or all the way around its margin. Contraction furrows may likewise be present. The major absence of iris diaphragm exposes the margin (equator) of the lens to view as a white line outlined by internal reflection within the lens. The fibrillary-like structures of the zonule are frequently seen making their attachment to the lens just anterior to this area. Frequently, on following the direction of the zonular fibrillae toward the ciliary body, the summits of the ciliary processes can be distinguished.

The interesting things about this anomaly are the defects found associated with it. Although cases have been seen with an otherwise

normal anterior segment, in most instances it is accompanied by other deformities, involving the cornea, chamber angle and frequently the choroid, retina, and optic nerve. In all the cases seen by me the corneal diameter was less than normal. Frequently a peripheral anterior synechia resulting from adhesion of the iris stump to the posterior corneal surface leads to glaucoma. However, it should be emphasized that glaucoma does not occur in every case. Lens defects varying from ectopia to microphakia have been found. Anterior or posterior polar cataracts, coronary, and lamellar opacities are common. In affected siblings aniridia may appear in one individual, while the associated defects may occur in others without aniridia. However, in the latter complete or incomplete colobomas may be present.

In most cases of aniridia the fovea is absent, and as a result there is lowered acuity of vision and nystagmus. Absence of foveal and macular reflexes can best be demonstrated in examination of the fundus by means of red-free light (Vogt). The simultaneous affection of the iris (pigment) and the fovea resembles another condition, namely, albinism, in which this also occurs. Both conditions result as a consequence of similar germinal defects. Vogt believes that genetically it is not surprising to find the absent or defective iris pigment linked with retinal (foveal) abnormality since the genes governing these tissues may be in the same or neighboring chromosomes. In many cases, despite the enlarged pupil, photophobia may be absent or minimal.

Today most embryologists believe that aniridia results from a primary ectodermal failure rather than from an initiating mesodermal aberration.

ANOMALIES OF PIGMENTATION IN THE IRIS

Iris in Albinism. The eye is affected in albinism, either as part of a universal (totally absent) or incomplete universal (relatively poor but not totally absent) congenital deficiency of pigment (hair, skin, or eyes), or it may be limited to the eye alone (*solum bulbi*) in which

case the general pigmentation, e.g., hair and skin, is normal.* Cases of solum bulbi in albinism have been described in which the iris or fundus alone were involved. However, even in those having an apparently normal fundus pigmentation, the macula has never been absolutely normal. In other words, as far as is known, albinism is always associated with a defective macula.

The presence or degree of associated secondary findings, such as underdevelopment of macula, nystagmus, head nodding, or photophobia, depends on or is coincident with the extent of the pigment deficiency or alterations in the retinal pigmented epithelium. Evidently perfect or complete differentiation of the retinal and choroidal layers depends in part on the presence of pigment but the exact *modus operandi* of these processes is still unknown.

On the other hand, Mann states: "A normally pigmented individual with coloured irides may show absence of foveal reflex and nystagmus with a normal choroid or with deficient pigmentation of the choroid only. It therefore seems that differentiation of the fovea and pigmentation are related not so much as effect and cause but as two characters which tend to vary together, possibly on account of juxtaposition of the genes."

Clinically, the appearance of the irides in these cases varies considerably, especially with the manner in which they are illuminated. By direct focal illumination or even diffuse light the color may vary from a light gray (almost white) to a yellow or golden brown. Sur-

* Mann, following Waardenburg, who attempted to bring order to the chaotic variations observed, classified albinism into seven types, as follows: (1) Universal albinism. The individual is totally deficient in pigment and shows underdevelopment of the macula and secondary nystagmus. The inheritance is that of a simple recessive. (2) Incomplete universal albinism. The patient is poor in pigment though this is not totally absent. The macula is usually underdeveloped. There may be head nodding, but nystagmus does not occur in all cases. The condition is a simple genetic recessive or an irregular dominant. (3) Isolated eye or fundus albinism. The general pigmentation is normal, the eye alone being deficient. Sometimes the iris is involved; sometimes only the fundus. The macula is underdeveloped. There may or may not be nystagmus and head nodding. The condition is a sex-linked recessive. (4) Aplasia or hypoplasia of the macula with or without deficient fundus pigmentation. This type behaves as a simple recessive, a sex-linked recessive, or an irregular dominant. (5) Aplasia or hypoplasia of the macula and the periphery of the retina with night blindness and diminution of the visual field. This is a simple recessive. (6) Nystagmus with high myopia and poor visual acuity (probably defective macula) but with normal pigmentation. This is a dominant. (7) Primary hereditary nystagmus without defective macula or albinism. This may be a recessive, a sex-linked recessive, or an irregular dominant.

PLATE XLVI

FIG. 1. Albinism—brown iris.

FIG. 2. Albinism—blue iris.

FIG. 3. Melanosis oculi—right eye.

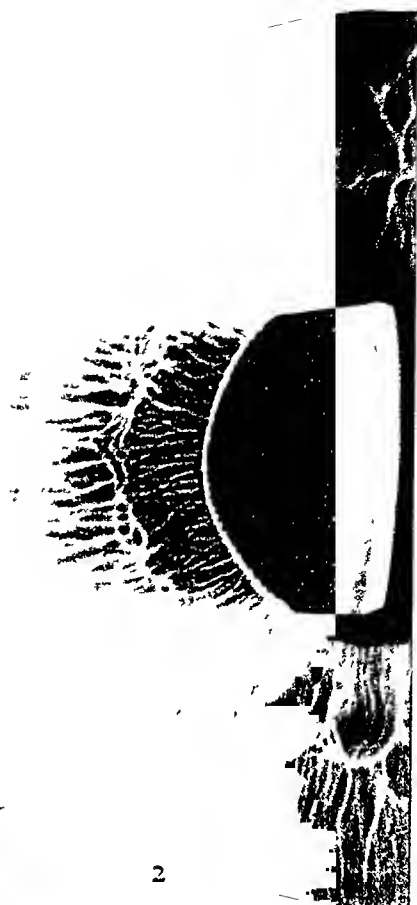
FIG. 4. Unaffected left eye. Same case as Figure 3.

FIG. 5. Sector of iris melanosis. (Diffuse illumination; high power.) Note the stars.

FIG. 6. Complete coloboma of the iris. Zonular fibers are seen in the area of the defect.



1



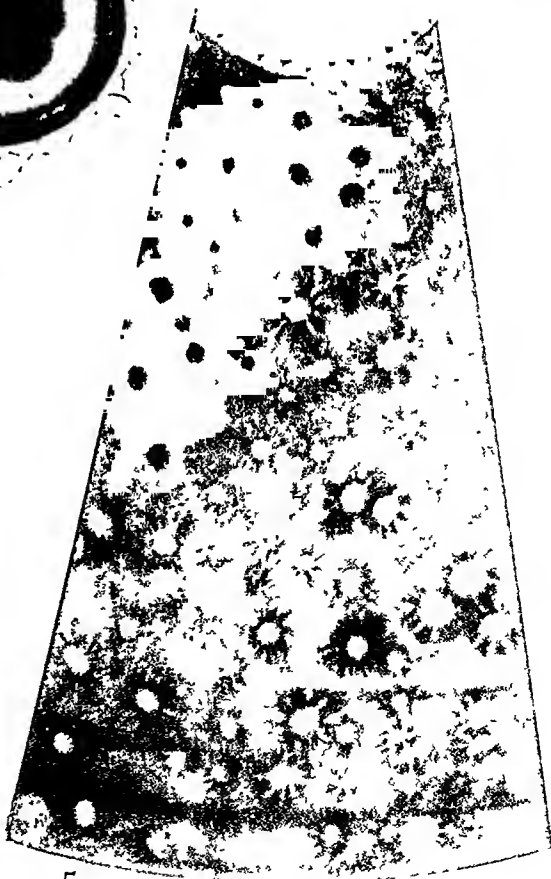
2



3



4



5

face melanophores in the iris may develop in an albino during childhood; in many normal individuals a blue or gray iris changes in color owing to the development of melanophores as late as 8 years of age. Whatever the actual hue of the albinotic iris by diffuse illumination, retro-illumination or transscleral illumination reveals features that are unmistakable, since the presence of a moderate amount of melanophores in the anterior border layer of the iris is not obstructive. It is the absence or deficiency of retinal pigment in the posterior iris layer that permits almost perfect transmission of light through the stroma by retro-illumination (Plate XLVI, figs. 1, 2). The reflected light from the lens outlines the trabeculae and the vessels. Fine superficial reddish vessels are frequently seen paralleling the radial trabeculae. These are also visible by direct focal illumination which, however, does not bring out the thicker walled, deeply situated vessels. On the other hand, transillumination of the latter gives the stroma a reddish hue. Also, by retro-illumination, deficiency of the retinal pigment frequently results in the appearance of radial light streaks, peripheral to the lesser circle. These streaks may represent the retro-illuminated system of structural furrows (see page 735). Likewise, when the pupil is partially dilated, a series of closely placed radial light spaces or lines can be seen. These spaces or lines probably outline the fine shallow grooves in the pigment epithelium, normally found on the posterior surface of the iris, extending from the pigment excrescences peripherally for a short distance. When the pupil is small, these lines are so close to one another that it is no longer possible to distinguish them separately. Vogt has described a dark semilunar line seen in retro-illuminated albinotic irides, which runs concentrically with the pupil and changes its location or moves according to the direction of the beam. Consequently, he believes that this line is not due to any structural change in the iris but results from an optic phenomenon (reflection and refraction) from the edge of the lens. This line has also been observed in cases of iritis or iridocyclitis when extensive and progressive depigmentation has occurred.

The typical ("rabbit's pupil") reflex of the pupil seen in ordinary

daylight illumination, owing to the absence or deficiency of pigmentation in iris and fundus and the consequent unobstructed passage of light through the iris and sclera, causes a pink or red glow to be seen by the observer. The red color of the glow in the pupil is due to reflection from blood in the vessels of the retina and choroid. This phenomena is accentuated by the focal beam.

Melanosis Iridis. Hyperpigmentation of the iris (true melanosiris) occurs in melanosiris bulbi, a condition characterized by increased pigmentation of the uveal tract, sclera, and occasionally of the optic nerve papilla. Its occurrence in fair or lightly pigmented individuals is rare.

Since this condition is preponderantly unilateral (90 per cent of cases) it offers little difficulty in diagnosis. As it occurs normally in pigmented races, melanosiris, if present, is found at birth. The scleral pigmentation in the form of slate-blue irregular patches is distinctive.* As a rule the fellow eye tends to be hazel (lightly pigmented) in color. However, in rare cases it may be blue or gray, so that a relative heterochromia iridis may result. The area of hyperpigmentation may be limited to a sector of the iris surface or it may be so extensive as to involve its entire anterior face. The affected area always appears darker by diffuse illumination (especially by daylight) than it does by direct focal or by intense afocal or diffuse illumination afforded by the biomicroscope. With the latter method of examination the overpigmented parts may even appear yellowish. The point to be emphasized is the density and profuseness of the pigmentation which approaches that of the normal spongy type seen normally in the Negroid iris. The layer of dense pigmentation may obscure the surface markings and the trabeculae outlines, giving the iris a thickened, smooth, and velvety appearance. However, with higher power, in most instances a system of peculiarly characteristic starlike formations is distinctive and is not found either normally in any race or in any pathologic condition (Plate XLVI, figs. 1, 2).

These stars or verrucosities are only seen in the area of hyperpigmentation and are generally more numerous distal to the lesser circle.

* See Volume I, page 155.

In some cases groups of closely packed perfect starlike figures are formed, having from 6 to 13 radiating projections (Koby), while in others the center of the verrucosity may resemble a tiny round mound, surrounded by a narrow dark ring from which short lines radiate. As a rule the centers of these formations are lighter in color than the hyperpigmented background and hence stand out almost whitish by comparison. In a way, they might be likened to the reflections of a star sapphire. No adequate histologic study of these structures has yet fully explained their clinical appearance, and consequently they are still regarded as resulting from an aggregation of melanophores. Reese (1925)⁵⁷¹ found groups of condensed and heavily pigmented melanophores.

Chapter Twenty

INFLAMMATION OF THE IRIS

THE iris, considered as the most anterior extension of the uvea, receives its blood supply chiefly from the long posterior ciliary vessels. These vessels give off no branches until they reach the posterior portion of the ciliary body. At this point, together with perforating branches of the anterior ciliary arteries, recurrent or retrograde branches supply the anterior portions of the choroid, while anterior branches go forward to form the *circulus iridis majoris*. The ciliary processes and iris receive their supply from this major circle. The venous drainage of anterior parts of the uveal tract (anterior choroid, ciliary body, and iris) and Schlemm's canal ultimately pools in the vortical veins. The high degree of vascularity of the uvea makes it vulnerable to the noxious effects of circulating toxins, viruses, or bacteria.

Because of a common nutrient supply and drainage system it is understandable that an inflammatory involvement, even though at first localized in one part of the anterior uvea, e.g., the iris, will extend by continuity or contiguity to the other parts. Even in cases of so-called pure cyclitis in which the major sign of iritis (*synechiae*) does not occur, indications of changes in the iris (atrophy) may be seen with the biomicroscope later during the course of the disease. Similarly there is definite proof that iritis is always associated with some degree of cyclitis and to a lesser extent with anterior choroiditis, and therefore is better termed anterior uveitis. Unfortunately there are no means at our disposal to examine the living ciliary organ directly; hence we can only infer the presence of inflammation in this body by the presence of exudative cells in the anterior chamber or vitreous. The rest has been gleaned histologically from eyes for

the most part so badly damaged as to have required enucleation. However, in the cases of inflammations of the iris, the biomicroscope is of incalculable value since it affords a better means for its direct observation and permits us to detect disturbances not only at the onset but also to observe them through their entire course to the final evaluation of the damage sustained. One should like to be able to state categorically that by biomicroscopic examination alone it is possible in every case to make a positive differential diagnosis, but in certain instances a differential diagnosis may require a survey that includes the employment of every diagnostic method known to medical science. The mere faculty of being able better to visualize the morphologic aspects by biomicroscopy has immeasurably added to our ability to make a differential diagnosis. For example: in an acutely inflamed eye, even from the very beginning interstitial keratitis can be differentiated by the corneal changes. In inflammatory glaucoma, the type of injection, condition of the cornea, size of the pupil, and depth of the anterior chamber quickly suggest that the intra-ocular pressure should be measured. Also an acutely developing shallowness of the anterior chamber may indicate the presence of an intra-ocular tumor. In my opinion, the ability to detect the presence of intra-ocular involvement by means of the aqueous flare is in itself one of the foremost diagnostic achievements. It is not within the province of this work to go into an extensive discussion of the etiologic classification of inflammatory diseases of the iris.* According to Woods, "Endogenous uveitis may con-

* The reader is referred to Duke-Elder's splendid chapter on diseases of the uveal tract in Volume III.¹⁰² Under the heading of the general etiology of uveitis he classifies them as follows:

1. Exogenous infections, derived from without, induced by (a) organismal infection, through a perforating injury or nick of the cornea or sclera (wound infection), (b) intra-ocular parasites, and (c) poisonous chemical, animal, or vegetable material.

2. Secondary infections affecting the uveal tract by continuity, or arising from some intra-ocular event.

3. Endogenous infection, the uveal tract being affected primarily from some cause operating elsewhere in the body.

In summarizing the relative frequency of the causes of endogenous uveitis (among adults in Great Britain) Duke-Elder rates focal infections (teeth, tonsils, and prostate) as first; tuberculosis (focal type) second; syphilis, third; metastatic lesions, fourth; acute infections and exanthemas, fifth; and skin diseases, sixth; while in a large residuum of cases the etiology is obscure and undetermined. However, it is doubtful whether in the present state of our knowledge such a classification is entirely valid. Continental European writers still hold that tuberculosis is the main cause of endogenous uveitis.

veniently be divided into three different groups: (1) sympathetic ophthalmia and the so-called endophthalmitis phaco-anaphylactica; (2) the various metastatic purulent infections of the eye which arise from direct infection by blood-born bacteria; (3) the usual non-purulent uveitis, iritis, or choroiditis, which so often is recurrent, and comprises the greater percentage of all uveal disease."

Woods states further: "The older writers divided disease of the anterior uvea into the 'serous' and 'plastic' iritis. To my mind a much better terminology is 'nongranulomatous' for these terms give an indication of the underlying pathologic process. In some instances it may be difficult or even impossible to make this differentiation, but as a rule the clinical pictures as well as the underlying pathology of these two forms of inflammation are quite different. The salient characteristic of these forms of uveitis are as follows:

"Non Granulomatous Uveitis. In the anterior uvea the onset is usually acute, rather than insidious, and ciliary congestion is marked with pronounced photophobia and lachrymation. The inflammatory reaction in the iris is usually slight and is limited to loss of luster, blurring of the iris pattern, and dilation of the capillaries. There are no nodules and but little tendency to the formation of posterior synechiae unless there are repeated recurrences. The aqueous ray is usually intense due to the outpouring of serum and there may even be a heavy gelatinous fibrinous exudate in the anterior chamber. The deposits on the posterior surface of the cornea are small and pinpoint, and are composed chiefly of lymphocytes. Heavy, greasy exudates do not occur, and there is little or no tendency to capsular clouding of the lens. Koeppe nodules are never observed. The course of a nongranulomatous iritis is usually short and the eyes usually recover with amazingly few residua. Only after repeated attacks is organic damage done to the eye.

"Granulomatous Uveitis. In the anterior uvea, the onset of the granulomatous iritis is usually insidious. The cellular reaction in the tissues is greater than the vascular reaction, and the ciliary congestion is not usually severe. Organic changes take place in the iris with thickening of the stroma from cellular infiltration with blurring of

the iris pattern and loss of the normal luster. Well defined nodules or tubercles on the surface of the iris, or diffuse localized thickenings suggesting nodules deep in the iris stroma, are sometimes present. There is a marked tendency to the formation of posterior synechiae, with greasy exudates on the anterior capsule of the lens and capsular clouding. The keratic precipitates are of the so-called 'mutton-fat' variety, and histologically consist chiefly of epithelioid cells. Koeppe nodules, or accumulations of epithelioid cells at the pupillary border of the iris, are not uncommon. The aqueous ray may be intense, but more often is rather mild."

Also in discussing iritis as an entity, one cannot always disassociate the concomitant ciliary involvement and during the acute phase the effects of any increase in intra-ocular pressure. From the viewpoint of biomicroscopy we shall limit ourselves to certain more or less chronologic or episodic inflammatory reactions occurring in the iris and which in most cases are not directly related to any specific etiology. However, it must be admitted that at times, certain well-defined patterns do appear, even in iritis, arising from endogenous causes, which will provisionally at least permit us to venture an etiologic diagnosis on clinical appearance alone, e.g., the nodules in tuberculous iritis, the hemorrhages and gelatinous exudates in gonorrhea, and circumscribed lesions in herpes, the luetic roseolae of the iris, etc. Consequently in such instances special descriptions will be given.

Naturally, the time of appearance of the lesions seen with the biomicroscope will depend on the type and the acuteness of the process. In certain fulminating forms the eye from the start may be so overwhelmed by the inflammation that edema and exudation, especially as they affect the cornea, make it impossible, except perhaps at the very outset, to observe any orderly sequence of events as they occur in the anterior chamber and iris. This is especially true not only in severe secondary forms, e.g., those secondary to infection in other parts of the eye such as the cornea (keratitis or serpiginous ulcer with hypopyon), lens, or vitreous body, but also in the severe primary types, e.g., acute hemorrhagic iritis or with the fulminating

acute purulent infections following a perforating wound or metastatic extension due to a bacteriemia in meningitis or pneumonia. These, from the beginning, may simultaneously involve part or all of the uvea, and depending upon successful localization of the process to the anterior or posterior segment may result in partial or complete obliteration of the anterior chamber in the former or to the formation of a vitreous abscess in the latter. In both, depending on the damage, the attendant sequelae may result in destruction of the eye through glaucoma or endophthalmitis. A rapid generalized purulent extension may result in a devastating panophthalmitis.

Frequently what starts out as a localized primary endogenous iritis may by extension develop into a serious form, either acutely, subacutely, or chronically resulting in endophthalmitis, secondary glaucoma, and ultimately an atrophic globe. This is the picture frequently seen in the granulomatous variety, particularly in tuberculosis.

The so-called chronic iritis usually is the result of minimal recurrent acute attacks, punctuated by periods of quiescence. The condition may remain localized to the anterior uvea (iris and ciliary body) for a long period of time before it affects deeper parts of the eye, or it may heal and remain stationary. Although the inflammatory symptoms (congestion and pain) may be slight, in the end great iris tissue damage from organization of exudates (adhesions), nodules, and atrophy results. Synechiae, anterior peripheral as well as posterior, lead to glaucoma — while the passage of toxins into the ocular fluids leads to changes in the lens and retina and ultimately to atrophy of the eyeball. The corneal involvement in iritis has already been referred to in Vol. I. Depending on the severity, chronicity and changes in intra-ocular tension, variations from transient corneal edema to permanent corneal opacifications may occur. In the same manner, all degrees of scleritis and episcleritis may at times develop. Chronic iridocyclitis leads to secondary disease of the lens and cataract formation. This may be initiated by affections of the capsular epithelium, evidenced by the not too infrequent appearance of anterior subcapsular cataract. Hyperemia of the iris and ciliary

body, together with exudation, causes variations in the intra-ocular tension. Not only does the quality of the aqueous (albuminous content) change but also its quantity. In acute cases reabsorption of the inflammatory products occurs quickly and there may be no changes in intra-ocular pressure. However, with excessive amounts of exudation, especially with recurrent or long standing processes, blockage of the drainage system results in increased intra-ocular pressure. In such cases the chamber may actually be deepened. But as secondary pathologic changes develop in the angle, e.g., swelling of the iris root or the extension of granulomatous processes from the iris or ciliary body, anterior peripheral synechiae lead to a shallow anterior chamber. The early stages of cyclitis on the other hand may even be accompanied by hypotony. Although the aqueous is highly plasmoid, slowing of the circulation in the ciliary body can result in a reduction in the quantity of aqueous. If such a condition endures for a long time (prolonged hypotony), atrophy of the globe ensues.

The granulomatous infections, e.g., tuberculosis, lymphogranuloma venereum, Boeck's sarcoid, brucellosis, lues, sympathetic ophthalmitis, perhaps special fungus and virus infections, are the chief etiologic agents in these complicated types of iridocyclitis. However, even these may cause only mild or abortive attacks.

Most cases of acute endogenous (nongranulomatous) iritis run a milder course and have a favorable prognosis. Depending on the intensity of involvement together with the natural resistance of the individual, its duration may vary from one to ten weeks. Reabsorption may be so complete that except for an occasional adhesion (posterior synechiae) or a pigment clump on the anterior lens capsule, no residual evidence of iris inflammation may be found. As a rule it is only in untreated cases of this type that a *seclusio pupillae* with its attendant sequela (glaucoma) results.

In general, the biomicroscope will reveal features that are of the greatest diagnostic import, especially at the very onset of iritis or iridocyclitis, and particularly in the nonsuppurative exudative types. The latter include the specific or nonspecific acute, subacute or

chronic forms, usually of endogenous etiology; in these forms the iris itself is nearly always accessible to biomicroscopic inspection throughout their course. In discussing exudative inflammations of the anterior uvea Duke-Elder ⁴⁰⁴ aptly states “. . . it is to be remembered that although many of these inflammations are called acute — and rightly so for their symptomatology and course may be exceedingly acute — in the pathological sense this, too, is an inaccuracy, for in contradistinction to the suppurative processes, the exudative inflammations of the uvea are all characterized by the mononuclear infiltration associated with subacute and chronic processes.”

The pathologic changes of the iris have been classified as vascular (hyperemia), exudative, pigmentary and atrophic. Generally at the earliest stages, from the standpoint of biomicroscopy of iritis, exudative changes are a more reliable and prominent feature than the others. For example, in brown irides congestion of the vessels indicative of hyperemia, one of the earliest changes in inflammation, may not be visible. Consequently, at the onset of an acute iritis the presence of exudation is a *sine qua non*. Exudation is recognized by the presence of an aqueous flare, cells in the anterior, posterior, or vitreous chambers, keratic precipitates,* efflorescences at the pupillary pigment margin (ectodermal) which when posterior organize and may form synechiae and efflorescences and nodules within or on the stroma (mesodermal) itself away from the pupillary margin.†

HYPEREMIA

Ciliary Injection. As early and striking objective symptoms of acute iritis, visible hyperemia occurs as pericorneal or ciliary injection. The so-called ciliary injection results from a congestion of the anastomosing conjunctival,‡ episcleral and scleral vessels, which are derived from the anterior ciliary vessels. Depending on the severity and extent of involvement, gradations of congestion vary

* See Volume I.

† It is well known histologically that nodules deep within the stroma and even those situated away from the pupillary margin may cause extensive posterior synechiae by extending or rupturing into or through the posterior pigment surface of the iris.

‡ Extreme congestion of the conjunctiva may be accompanied by chemosis and even redness and swelling of the upper eyelid.

from a faint pink when slight to a deep purple when the deeper vessels are involved. As a rule the injection encircles the cornea but at times it may be localized to a sector corresponding to the area of greatest iris involvement. For example, the location or point of maximum intensity may correspond to a place where nodules are found. It is also possible that the normal anatomic distribution of the deeper branches of the anterior ciliary vessels may also be a factor in determining where the injection will be most marked. Just as hyperemia and ciliary injection indicate inflammatory reaction, subsidence of this congestion naturally suggests regression.

Hyperemia of Iris Vessels. Although most of the radial fibers of the superficial iris layers carry blood during fetal life, this function ceases just before birth. We see these structures as the ordinary iris trabecular meshwork unless they are obscured by melanophores, e.g., in the brown iris. With the exception of albinism, blood-carrying trabeculae are rarer than those without vessels. Even in a normal blue or gray iris, only occasionally is a radial or angulated vascular loop found (Plate XLI, fig. 6). These vessels (probably veins) are either free and unsupported or are bordered by a delicate, whitish sheath, giving the impression that they occupy the central part of a fine trabecula. At times they may branch and join up with a vessel of another sector. I saw such a vessel in a case in which there was a slight ectropion of the pigment border of the pupil. This vessel, after leaving the periphery, branched in three directions and in extent could be traced over half the iris surface. Both the vessel and ectropion occurred in the same iris sector below and temporally. There were no indications of an inflammatory cause.

Because of their thick opaque walls, the large, more deeply situated vessels are not visible as such biomicroscopically. They may be discernible only after marked depigmentation and extensive atrophy of the iris substance, e.g., *rubeosis iridis glaucomatosa*. From the pathologic standpoint, an increase in number and prominence of iridic vessels bespeaks either iritis or venous stasis. The latter is seen in diabetes (rubeosis), glaucoma, thrombosis of the retinal veins, and

in intra-ocular tumors (including hemangiomas of the iris) (Plate LI, figs. 1, 2, 3, 4).

Biomicroscopically, in acute iritis, hyperemia is manifested by the presence of dilated vessels, which as a rule have a radial direction. Such vessels are best seen in the ciliary zone and tend to branch as they approach the lesser circle and pupillary zone. Vessels may be found in several or in a single iris sector, either paralleling the whitish trabeculae or crossing them at various levels in the stroma. Usually they are only seen in blue or gray irides since the heavy obscuring surface pigmentation of the brown iris prevents their detection. Hyperemia of the iris vessels undoubtedly causes some of the color changes seen in inflammation. The addition of red color (from the vessels or from small hemorrhages) to a blue or gray iris will cause a greenish appearance. A light brown iris will turn a reddish-brown. The discoloring effect of hyperemia will, as just mentioned, affect the dark brown iris least. The fact that only a few trabeculae carry visible vessels, even in severe iritis, indicates that for the most part they completely lose their original blood-carrying function. At the onset of an acute iritis (in light colored irides, blue or gray) the pre-existing vessels become more visible or previously empty vessel-bearing trabeculae develop a reddish central line indicating the presence of blood. In other cases vessels with no discernible sheath may be seen. These tend to appear in the neighborhood of the collarette or in or near the sphincter zone. Here they may run circularly (concentric to the pupil) or assume a congery of twisted formations. When of fine or delicate outline, the frill (lesser circle) not infrequently will contain blood. In this case the outlines of its arcade-like structure become apparent, forming a vascular arc or complete circle circumferential to the pupil. In others small local congeries of vessels (see roseolae and rubeosis, Plate LI) may appear in one or more places in the region of the frill; at times they are irregularly distributed in the ciliary zone. Most of these visibly dilated vessels in cases of simple iritis return to their previous blood-free state as the process abates. But at times such a vessel retains its red color permanently. Recurrent attacks of chronic iritis favor persistence of newly formed vessels.

As a result of prolonged or severe stasis, perhaps together with atrophy of the stroma, iridic vessels may tend to lose their delicate trabecular sheath, and as is the case with new formed vessels, stand out in bold relief to form the picture known as rubeosis iridis (Plate LI, figs. 1, 2, 3, 4). The presence of such vessels in the iris periphery and in the chamber angle leads to the formation of anterior peripheral synechiae and inevitably to secondary glaucoma. In severe exudative granulomatous infections, e.g., tuberculosis of the iris, new vessels may not only invade the iris but may proliferate irregularly over the anterior lens capsule especially when a pupillary exudate is present (Plates XLIX, fig. 7; LI, fig. 7). Pathologically, hyperemia is one of the first reactions to tissue insult. But from the biomicroscopic standpoint, since its presence in the iris may escape detection, e.g., as in a brown iris, one acknowledges this sign as a definite indication of iritic irritation or venous stasis; but for the earliest proof of onset of iritis; one relies more on the presence of exudate (increased aqueous flare with or without cells or debris) and corneal edema (epithelial and endothelial). I have verified this point many times at the onset of iritis in cases of gray or blue irides in which an outstanding aqueous flare with or without floating cells preceded by days an increase in the number of visible vessels.

In special cases of acute iritis (e.g., gonorrheal and herpetic iritis) hyperemia of the iris itself may be so marked as to constitute an outstanding feature. The extensive vascular engorgement may lead to formation of visible groups of vessels and petechial hemorrhages, and eventually to hyphemia of the anterior chamber (Plate LII, fig. 2). An instance of this was seen in a case of acute iritis occurring in an eye, which five years previously had been trephined for chronic simple glaucoma. Up to the time of the iritis the eye had been entirely free from inflammation, the intra-ocular pressure being stabilized at 12 mm. Hg (Schiötz). With no previous history of conjunctivitis, the patient suddenly developed pain and redness (pericorneal injection) in this eye. On the following day biomicroscopy revealed a distinct flare in the aqueous, containing fibrinous exudate and pigment cells. The miotic pupil was bound down by an exudative membrane in its lower parts. There was no exudate

seen in the operative coloboma above. There were many radiating vessels in the iris stroma visible as reddish lines. However, by indirect light in the pupillary zone and near the lesser circle small bright superficial spots (hemorrhages) were seen. Although predominantly they were situated near the frill, several isolated ones were found in the ciliary zone. A few days later a hyphemia suddenly developed and the eye became hard, extremely congested, and painful. The eyelids were chemotic, the eye was protuberant and had limited adduction. After a few days enucleation became imperative.

Edema. Edema or swelling of the iris tissue itself, with sogginess, obscuration of the delicate trabecular architecture and change in color, is manifested only after the process is well established. Naturally the time of appearance of such changes varies from case to case and depends on the violence or duration of the inflammation. Many writers, in describing the earliest changes of iritis, speak of a variable haziness of the iris surface tissue. Some even imply that they received the impression that a thin veil was present over the iris surface but that this veiling never obscured the trabecular structure. Edema of the iris is difficult to evaluate in the early diagnosis of iritis. I have examined innumerable cases in the earliest stages of inflammation and as yet am not convinced that it is possible to diagnose the presence of early edema. Both the veiling effects of the cornea (which may be more responsive) and an increase in aqueous turbidity interfere.

Exudation. Exudation occurs in iritis not only in the chambers of the eye and on their boundaries but also on and within the iris itself.

EXUDATION IN THE ANTERIOR CHAMBER

*Aqueous Flares.** Exudation in the form of albumen in the aqueous, with or without the presence of cells, results in the production of a visible aqueous flare (Tyndall phenomenon). This is a cardinal finding at the very beginning of iritis, cyclitis, or anterior choroiditis and to my mind its early detection is one of the most important diagnostic advances afforded by biomicroscopy. Whether

* See Chapter 16 on anterior chamber, Volume I.

or not dilatation of pupil retards the amount of exudation is still a moot point. Gilbert⁴⁵⁰ described a case of iritis, which showed only a few efflorescences in front of and behind the pupillary seam superiorly. The picture remained unchanged for a week—but when the patient discontinued treatment for a few days, there was a marked increase of exudate. At this time the upper part of the pupil was covered and bound down by the extensive exudate. With renewed atropinization the adhesion separated, and further exudation ceased, so that in a few days the picture resembled that of the earlier state.

Patients frequently seek medical attention because of a slight blush of conjunctival injection. The presence of even a minimal aqueous flare at this early stage warns of an intra-ocular process and dispels any idea that the condition is merely a simple conjunctivitis—an error frequently made in prebiomicroscopic days.

As the beam traverses the normal anterior chamber, a dark optically empty area, representing the depth of the anterior chamber, occupies the space between the relucant corneal and lenticular blocks. At the point where the protein content of the aqueous increases (due to exudation) to at least ten times that of the normal, the beam becomes visible as a grayish flare as it passes through the aqueous.* The possibility of a physiologic flare in the normal eye, owing to the slight albuminous content of the aqueous, can be disregarded because of its delicacy and because it is only seen under special conditions. Practically, a visible flare should always be considered as a pathologic manifestation.

The intensity of the flare increases with the degree of aqueous turbidity, so that if one directs the focused beam through a highly turbid aqueous and observes it with the unaided eye, no interruption of the Tyndall phenomenon will be seen from the surface of the cornea to the posterior surface of the lens. In most instances, even when the condition is observed at an early stage, floating cells, indicated by moving points in the flare, will be observed. To see these, higher magnifications of 20 × or more are required.

At the onset, in most cases of uveitis that I have studied the cellu-

* See Volume I.

lar elements were unpigmented and appeared as grayish dots. On the other hand I have frequently observed that from the very beginning of an iritic attack, even in a blue or gray iris, the cells were pigmented. It is my impression that pigmented cells are more liable to be seen when the inflammatory processes were severe and fulminating. They were common in inflammatory glaucoma. This also held true for keratic precipitates appearing later. However, it should be emphasized that at the present time no definite etiologic or pathognomonic significance can be attached to the color of cells *seen in these inflammatory conditions. As a rule the tendency to form large keratic precipitates is more marked in cyclitis than in acute iritis, though exceptions are frequently seen. The appearance of a heavy corneal precipitation, e.g., "mutton fat" clumps, in acute iritis suggests accompanying cyclitis (Vol. I.).*

Subjectively the presence of an aqueous flare is always accompanied by blurring of vision. This symptom often precedes all others. As the intensity of the inflammatory processes mounts, increasing changes in the cornea will be observed.* These consist of endothelial bedewing, precipitates, folds in Descemet's membrane, edema of the stroma and epithelium. The summation of these alterations results in a gradual clouding or increase of relucency of the cornea. When this becomes marked, observation of the aqueous flare becomes difficult especially when higher magnifications are employed. In such cases it is preferable to disregard the microscope and to observe the

* Invariably, delicate changes are present in the endothelium almost at the very onset of uveal irritation. I had occasion to examine a physician, aged 36, who on awakening became aware of a slight blurring of vision. During the day he noticed a slight blush of the bulbar vessels in both eyes. Biomicroscopic examination revealed a slight increase in aqueous flare bilaterally. In each eye there was a faint stippling of the endothelium in the lower parts of the cornea. The irides appeared normal and except for minimal neuralgic pains about the eyes and the blurring, there were no symptoms. The fundi were normal. On the following day the blurring seemed less and the injection had cleared. After the fifth day the aqueous flare and cells were not visible and the delicate stippling (bedewing and keratic precipitates) of the endothelium in both eyes was absent. He received no treatment either locally to the eyes or systemically, except that he gave a history of having had a subsequent suspicious apical pulmonary lesion. He had had tuberculosis five years before but since then had been perfectly well. Five years later he developed hemoptysis and active pulmonary lesions were again found.

I have also seen minimal reactions of this type in association with a moderate rise in intra-ocular tension in a case which later proved to be one of chronic simple(?) glaucoma. Coinciding with the lowering of tension after the use of miotics the flare disappeared.

passage of the beam through the anterior chamber with the unaided eye, using the small cylindrical bundle or the slit of 0.5 mm. height. The smaller sized beam cuts down considerably on the interfering dispersion of light resulting from the increased corneal relucency and aids in the perception of the aqueous flare.

Cells in the Aqueous. The ability to detect a delicate increase in aqueous flare at the onset of uveal inflammation may tax the skill of the observer. But the presence of cells at this stage, in either the anterior chamber, retrolental space (anterior vitreous) is a sign of paramount diagnostic significance. Although cells in the anterior chamber can occur physiologically in special instances,* as a rule their presence is indicative of an intra-ocular inflammatory reaction. Comparison with the fellow eye, provided it is normal, usually dispels any doubts concerning this sign. I have seen cases in which the aqueous flare was very marked before any individual point-like dots (cells or pigment) were apparent, while in other cases, in which there was an almost completely imperceptible flare, from 5 to 20 cells could be counted, floating with the convection currents.

After a process becomes well established and the inflammatory reaction reaches its height, there will be no difficulty in verifying the presence of flare or cells, and when the well-marked aqueous flare is examined with higher power, it may be found to be composed of a densely packed stream of points. When the aqueous becomes highly "plasmoid," the usual convection currents cease. Resumption of cell movement is an early sign of diminution of the inflammation.†

As previously mentioned, in fulminating cases of iridocyclitis and in such conditions as inflammatory glaucoma, or acute interstitial keratitis, edema‡ and cloudiness of the cornea may hamper observation of the anterior chamber or retrolental space; but such hindrance may be overcome to a degree by cutting down on the size of the beam and by persistent observation over a period of time.

* See Volume I, page 434.

† See Volume I, page 568.

‡ A drop of glycerine placed on the cornea frequently will aid in a temporary clearing of epithelial edema.

TYPES OF CELLS AND EXUDATE

Pigment. As was previously indicated, in severe cases of anterior uveitis pigmented cells may be observed in the anterior chamber flare. These pigment granules probably come from the pigment excrescences of the pupillary margin (seam) or from the retinal pigment posteriorly; it is not known whether any of this pigment comes from broken-down melanophores during the acute phases of a slight or moderately mild iritis. Destruction of the melanophores probably depends on the location, intensity or chronicity of the iris involvement. In acute inflammatory glaucoma it is not unusual to find a great dispersal of iris pigment in the anterior chamber, on the posterior corneal surface, and anterior lens capsule as well. This may also be demonstrated in the iris after the acute phase has subsided by the appearance of depigmented or vitiligoid areas (Fig. 323; Plate LI, fig. 5). In chronic and dystrophic states, depigmentation of the iris stroma is commonly seen. Pigment granules are also generally found in the aqueous and as deposits on the chamber boundaries immediately after injury to, or following surgery of, the iris. Although pigment granules are found on the anterior capsule as congenital rests (remains of the tunica vasculosa lentis anterior and the retro-iridal lines), their presence on the posterior lens capsule, retrolental area or vitreous generally indicates an antecedent inflammatory process.

Erythrocytes will always be seen in the anterior chamber after hemorrhage. (See Plate XLIX, fig. 2; Vol. I, Plate XXXI, fig. 1.) Owing to massing of the cells, hyphema either in the anterior chamber, retrolental space, or vitreous appears bright red in color; whereas the individual red cells seen floating above it are distinctly yellowish in hue. This is especially true in hemorrhages of the vitreous after the cells have diffused through the gel. As time goes on the coloring matter in these cells may be washed or laked out; the points then become whitish. As a rule the deposits from hemorrhage are dustlike in character while those from inflammation are coarser. One should always be cautious in making deductions from

biomicroscopic appearance of delicate variations in color of cells seen in the aqueous and even more especially in the retrolental space or anterior vitreous.



FIG. 323. Vitiligoid area in iris near operative coloboma, in a case of chronic iritis and secondary glaucoma. Note pigment granules on the anterior lens capsule and scattered over the iris surface.

With higher magnifications errors resulting from refraction and aberration may cause false color changes. The color of a point in the retrolental space or vitreous may vary with the type of light source, type of illumination, or the magnification employed. As a rule, the stronger the illumination and the higher the magnification, the whiter a deposit will be. For example, the red color of blood is seen better macroscopically than microscopically. For these

reasons, when attempting to determine the color of cells, one is advised to observe the beam with the unaided eye or under very low magnification. It is necessary to point out that an admixture of blood cells in a pre-existing fibrinous exudate (iritis) gives the chamber and the iris itself a muddy appearance.

White cells. It is my impression that white cells predominate in cyclitis and in choroiditis. This also seems to be the case in iritis except in certain fulminating cases where the cells may be pigmented (yellow, brown or reddish) from the outset. However, during the first few days of developing iritis, the cells in the aqueous are whitish but become more pigmented as the process develops (phagocytosis of pigment granules). Consequently, in the very early stages of uveitis one cannot make a differential diagnosis by the color of the cells alone, or ascertain by this sign in which part of the anterior uvea the inflammatory focus predominates. It is generally assumed that the smallest white dots represent individual white cells and that the larger ones are clumps of white cells.

Fibrinous Exudates. In severe cases of iritis (e.g., gonorrheal iritis) the exudate may agglomerate and may easily be distinguished as dense clouds or even grossly solid masses. Large agglomerations of cells or products of inflammation may sink by gravity to the lower portions of the anterior chamber, e.g., hyphema, hypopyon, solid exudates in severe keratitis, or metastatic masses from neoplasms.

An outstanding example of iritis, in which filiform threads or larger coagula form in the anterior chamber, is the one caused by the gonococcus. Here, the anterior chamber may be entirely or partially occupied by a mass of gelatinous exudate, which may obscure the iris and simulate a dislocated lens (Fig. 324). It is interesting to note how quickly such a jelly-like mass may form and absorb. Within a week such an exudate can appear and disappear, leaving no visible keratic precipitates behind. On the other hand, the denser and more opaque massive exudates seen in the granulomatous infections tend to organize and to become vascularized, thus leading to dire sequelae.

Many authors have stressed the importance of discovering cells

in the anterior chamber, the retrolental space, and the vitreous in suspected sympathetic ophthalmitis. Butler^{33a} states it is one of the most valuable features of biomicroscopy "because it allows us to



FIG. 324. Gonococcal iritis. Large gelatinous mass in the anterior chamber. Two iritic adhesions.

recognize sympathetic ophthalmitis several days in advance of the appearance of the more obvious signs." In commenting on Vogt's dictum, that if the slit lamp had no other value than the ability to recognize sympathetic ophthalmitis at an early date, it would justify its cost and the trouble necessary to master its technique, Butler says, "We feel that all who have saved an eye by indications of the slit lamp will fully agree with him."

Parasites: Onchocerciasis. Through the courtesy of Dr. M. Puig Solanes, I had the unusual privilege of seeing two of his cases in which it was possible to observe microfilaria in the cornea and in the anterior chamber. In a personal communication he stated that corneal lesions are found in 85.7 per cent of onchocerciasis, and that it appears in three clinical forms: (1) as superficial punctate keratitis, (2) as an interstitial keratitis, and (3) as a vascular keratitis. Biomicroscopically it is possible to observe the microfilaria in

the parenchyma of the cornea and in the anterior chamber. In the cornea they can best be seen by retro-illumination. By this form of illumination they appear as elongated filaments of uniform size and

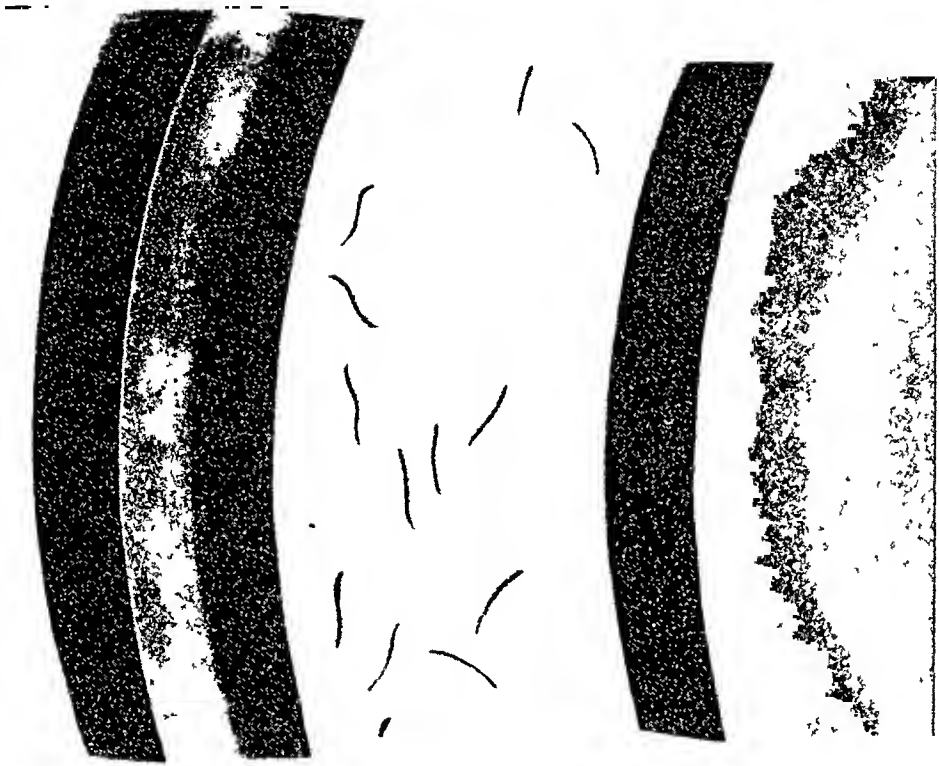


FIG. 325. Parasites (microfilaria) in the anterior chamber (onchocerciasis).

shape. In the anterior chamber they are easily seen in the aqueous flare (iritis generally being present) as thin curved lines, wriggling freely in the aqueous. The ability to recognize microfilaria in the cornea and anterior chamber is an important asset in the diagnosis of this disease (Fig. 325).

EXUDATION (DEBRIS AND CELLS) IN THE SO-CALLED RETROLENTAL SPACE *

When the nitra beam is sharply focused on the posterior lens capsule a dark intervening space (space of Berger) is seen between it and the vertically folded anterior face of the vitreous proper. When making observations in this area it is preferable to have the

* A more detailed consideration of the anatomy and biomicroscopic appearance of the retrolental space will be treated in the chapter on the vitreous, page 1375.

angle between the axis of observation and illumination as narrow as possible, and to use the cylindrical or short slit opening of the diaphragm with narrow width of aperture (optic section).



FIG. 326. Cyclitis. Cellular deposits on the posterior lens capsule, in the retrolental space and vitreous. A band of vitreous is adherent to the posterior lens capsule below.

For purposes of orientation it is advisable to begin observations with objectives of low magnifying power. The finest structures, such as particles and cells, unless obscured by haziness of the intervening optic media, are easily seen as a consequence of the intrinsic magnification induced by the dioptric power of the anterior media (cornea, aqueous, and lens). A cell will appear almost twice as large on the posterior lens capsule or in the retrolental space as it would in the anterior chamber (Fig. 326).

The stronger the illumination, the more reluctant Berger's space appears. With the arc illumination, or even with an overvolted nitra lamp, Berger's space, visualized as a dark, optically empty band

when ordinary nitra illumination is employed, now appears more reluctant (owing to an increase in the intensity of the Tyndall phenomenon). Not only does the fibrillar vitreous structure now appear to have approached closer to the lens but within the retrolental space itself is revealed a delicate fibrillary structure. This would indicate that the substance occupying the so-called retrolental space is vitreous (hyaloid *), or is derived from the vitreous rather than the aqueous. Hence, we have to consider that exudation in this region and on the posterior lens capsule is part of a general exudation into the vitreous. This is borne out by personal observation in several cases that inflammatory deposits on the posterior lens capsule† are either preceded or at least associated with exudation into the hyaloid vitreous (retrolental space) as well as in the deeper definitive vitreous.

It is impossible to state definitely whether, in inflammatory states of the uvea, exudates (cells or fibrin) appear in the retrolental space before they do in the anterior chamber. According to most writers, the reverse is true. However, in certain instances (sympathetic ophthalmia) it has been reported that depositions in the retrolental area and in the vitreous proper were not only the first to appear but were the only indications of a disturbance. It has been my impression that in iritis exudation of flare, cells, or both, is first seen in the anterior chamber, while in choroiditis (anterior and posterior) exudates (chiefly in the form of cells) are initially found in the retrolental space (vitreous) and on the posterior lens capsule when these areas are accessible to examination. As the inflammatory processes progress, inspection of the retrolental space may no longer be possible owing to corneal haziness and turbidity of the aqueous. Consequently it may be stated that in inflammatory conditions, examination of the retrolental space will be fruitful in the earliest stages or later during the period of regression when visualization in this area becomes possible; it is also most advantageous in cases of anterior choroiditis when the lesion is so far forward as to fall out of the range of the ophthalmoscope. In the latter instance the pres-

* See chapter on vitreous, page 1371.

† See paragraph on inflammatory deposits of posterior lens capsule, page 1217.

ence of cells or exudate in the aqueous as well as in the retrolental space (vitreous) or on the posterior lens capsule may be the only clue of an inflammatory process. The color of these cells may assist in differentiating between hemorrhage and inflammation. As previously described the color of blood cells will vary from bright red to yellow; uveal pigment appears reddish brown while ordinary exudative cells are grayish.

As in the anterior chamber where keratic precipitation occurs, exudates in the retrolental space and vitreous proper may be deposited on the posterior lens capsule. It should be remembered that the precipitation of inflammatory products on the posterior lens capsule ordinarily is rarer and much less in extent than that occurring on the posterior corneal surface. Generally speaking, deposits on the posterior corneal surface will be found earlier than those on the posterior lens capsule. Koby does not believe that a correlation can be established between the two. It can be stated only that a keratic precipitate occurs more frequently alone than in conjunction with deposits on the posterior lens capsule, rather than the converse.

Probably owing to the fact that convection currents behind the lens are absent or are not as marked as those in the anterior chamber (owing to the consistency of the retrolental substance and because there is little or no temperature gradient effective), the deposits on the posterior lens capsule tend to be more diffusely arranged than they are on the posterior surface of the cornea, e.g., an inverted V. Also one must consider the difference in curvature between that of the posterior lens surface (convex) and that of the posterior corneal surface (concave). The usual deposits on the posterior lens capsule are dotlike, star-shaped, or irregularly angulated. Pigment granules are very common and are frequently noted especially in association with a generalized vitreous pigmentation (e.g., following retinal separation). Further consideration of inflammatory deposits on the posterior capsule will be given on page 1217.

The fate of posterior capsular and retrolental (vitreous) deposits (exudate and pigment) vary. They may be resorbed and entirely disappear, or individual cellular elements (especially in the vitreous

proper) may remain present for long periods of time. When the exudation is massive (as may happen in some of the severe granulomatous or septic infections and in the case of uveal inflammations of infants) organization leads to the formation of dense membranes. Consequent to the formation of thick membranes are the sequelae of shrinkage and atrophy of the globe. Lens opacification frequently precludes biomicroscopic examination of such extensive changes.

Efflorescences and Nodules. Since the advent of the biomicroscope attention has been called with increasing frequency to the presence of exudative agglomerations in the form of efflorescences or nodules, in inflammatory conditions of the iris and ciliary body. These may occur not only at the pupillary border but also on or in the iris stroma itself, during acute, subacute, or chronic inflammations of the iris and ciliary body, and have even been known to form in cases in which there is no ciliary injection. These exudative lesions are usually not accompanied by any loss of tissue substance.

Histologically these formations range in composition from coagulated fibrin with or without cells (pellucid efflorescences) to the more solid cellular accumulations (nodules). Vogt found histologically that the nodules were composed of lymphocytes, especially large mononuclear cells, and epithelioid cells, with caseation. In his experiments, animal inoculation with fragments of iris containing nodules proved negative for tuberculosis, although there was involvement of lymph nodes. Biomicroscopically, efflorescences (especially those at the pupillary margins) came into prominence after they were described by Gilbert (1914)⁴⁴⁹ and by Koeppe (1917)⁵⁰⁴ and since then, those found at the pupillary border have been commonly known as "Koeppe nodules." The generalized term "nodule" for these bodies is in my opinion somewhat unfortunate since this term infers a solid structure. Consequently, I feel that the term "efflorescence" or "floccule" is more appropriate for the delicate and almost translucent formations and that the designation "nodule" should be reserved for those appearing more opaque and solid.

In the present state of our knowledge no specific etiologic significance can be attributed to efflorescences or nodes seen in iridocyclitis.

However, one is tempted clinically at least to venture certain observations concerning them. For example, the smaller ephemeral and translucent efflorescences, especially occurring at the pupillary margin (ectodermal) commonly occur in the acute nonspecific types of exudative iritis and iridocyclitis, whereas the larger more solid or opaque nodules in any location suggest the possibility of a granulomatous process. Excluding a specific etiology, e.g., such as syphilis, leprosy, sympathetic ophthalmitis or Boeck's sarcoid, European writers are prone to attribute a tuberculous etiology to most cases of iritis or iridocyclitis exhibiting nodes of the latter type, particularly when the process is chronic. Vogt has drawn an analogy between them and the skin tuberculides (Darier) of the extremities and back, and also to the efflorescences in phlyctenular keratoconjunctivitis. It may well be that European authors are correct in their assumption that most chronic forms of anterior uveitis which they see, especially those exhibiting nodules, are of tuberculous origin. However, this may not hold true for other countries or other localities. For example, recently it has been shown in this country that brucellosis (an infection that is more widespread than is generally suspected) is not an uncommon cause for recurrent iritis. Also the frequency of uveitis in Boeck's sarcoid has been noted. Both Heerfordt's disease (uveoparotid fever) and some cases of Mikulicz's syndrome may be manifestations of sarcoid. Iritis caused by these infections show no special features biomicroscopically that might distinguish them from tuberculosis.

From the standpoint of their histologic appearance and negative bacteriologic findings, the formation of iris nodes strongly suggests the idea that they are the result of an allergic response.* The lack of uniformity in the description and terminology of these structures is understandable since there is such a great variance in their clinical appearance.

Whatever their physical appearance, nodes have been classified according to their point of origin as ectodermic or mesodermic.

* One could be tempted to consider efflorescences (similar to phlyctenules) as a tuberculo-allergic response and the more solid ones (especially the intrastromal nodes) as actual tubercles.

PLATE XLVII

FIG. 1. Iritis. Keratic precipitates. Pigment deposits on the anterior lens capsule. Pigmented efflorescences.

FIG. 2. Efflorescence (early).

FIG. 3. Same efflorescence in Figure 2 seen later—pigmented.

FIG. 4. Efflorescences.

FIG. 5. Floccules of Busacca (luetic).

FIG. 6. Efflorescences.

FIG. 7. Efflorescence—high-power view of Figure 5.

FIG. 8. Broad efflorescences.

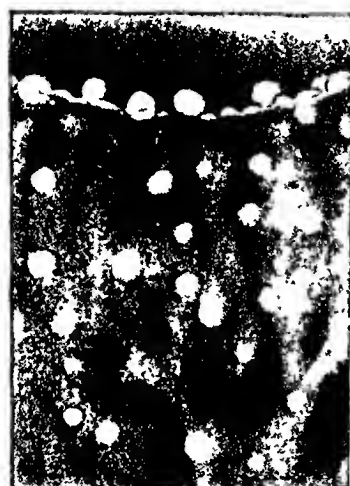
FIG. 9. High-power view of Figure 8 by direct focal illumination.

FIG. 10. High-power view of Figure 8 by retro-illumination.



2

3



5



6



8



9

10



Those arising from the retinal pigment layer, either on the excrescences of the pupillary margin (seam) or just behind them from the posterior surface of the iris, are called ectodermic. Those appearing on the iris stromal surface (flocules of Busacca) or within the stroma itself, are known as mesodermic.

Ectodermal Efflorescences and Nodules. Concomitant with the appearance of definite aqueous turbidity and keratic precipitates is the frequent finding of small whitish efflorescences at the pupillary border. These are located either directly on the intact pigmented excrescences or seemingly arise from the posterior surface of the iris to project themselves into the pupillary area sufficiently to be visible as small, soft, white, translucent fluffs (Fig. 327; Plate XLVII, fig. 2). At times their smooth rounded surfaces remind one of a vesicle. They are ephemeral, often disappearing within a few days to a week after their onset. Such smooth efflorescences are commonly seen in the early stages of luetic iritis.

Another type of small ectodermal efflorescence, frequently seen in nonspecific iridocyclitis, is the one whose consistency or contour is not unlike that of the large mutton-fat type of keratic precipitate. This form predominantly seems to arise from the posterior surface of the seam and usually is in contact with the anterior lens capsule (Plate XLVII, figs. 1, 7, 8, 9, 10). Although more anteriorly situated ones may lie on the excrescences, those not in contact with the lens capsule are less likely to form adhesions and they tend to disappear at a slower rate than they form. The surface of these lesions is rough and granular and tends, particularly in the case of brown irides, quickly to become dusted with pigment (Fig. 328). According to Meesmann's conception they are more often found on the lower parts of the pupillary border. For this reason, together with the fact that they are likely to be near the vertical axis, he believes that they are deposited in a way similar to keratic precipitates, namely by the action of the aqueous convection currents and gravity. However, it is generally held that these efflorescences arise *de novo* in the iris stroma. The more solid efflorescences tend to form posterior synechiae, especially if their posterior surface touches the anterior

lens capsule. If the pupil is dilated, one will frequently see a spot of pigment and exudate at the lens capsule at the original point of contact. In addition, occasionally one may observe a few dilated iris

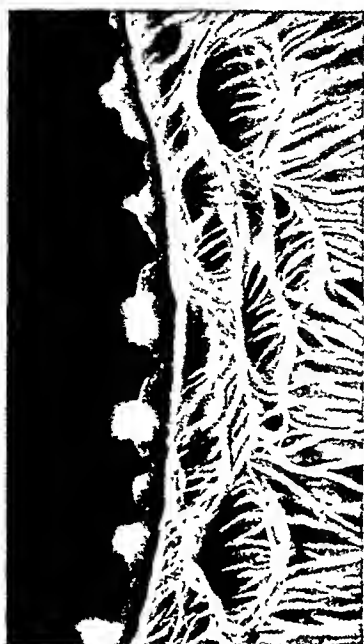


FIG. 327



FIG. 328

FIG. 327. Efflorescence at pupillary pigment border in early stage of iritis.
 FIG. 328. Well-formed efflorescence dusted with pigment at a later stage of iritis.

vessels in the neighborhood, although this occurs only at a late stage. However, it should be noted that not all of these efflorescences result in adhesions. The smaller, more anterior ones may slowly disappear leaving no trace. I recently observed a case of acute iritis of unknown etiology in which three grayish efflorescences were found at the lower pupillary margin (the iris was dark brown in color). In about a week they became so heavily pigmented that they no longer could have been identified as efflorescences had this change not been closely followed (Plate XLVII, figs. 2, 3). At this point, on dilatation of the pupil, they simulated the pigmented masses (synechiae) such as those seen adhering to the anterior lens capsule after rupture of a synechial adhesion. Koby was convinced that this form of efflorescence resulted from deposits of inflammatory cells after observing that they formed on a layer of pigment which remained adherent to the lens after iridectomy. Both Spicer and Vogt have observed

them on the anterior lens capsule. Meesmann found them in one case attached to threads of persistent pupillary membrane. In a case of exudative iridocyclitis I have seen similar bodies attached to freely floating strings of fibrinous material in the aqueous. (See Vol. I, Plate XL, fig. 2.)



FIG. 329. Nodule near the pupillary border (tuberculous iridocyclitis).

The larger, more solid appearing structures at the pupillary margin (which in my opinion are not ephemeral as efflorescences) really deserve the name "nodule" (Fig. 329). One cannot, however, be dogmatic concerning the duration of even the solid type of nodule at the pupillary border. Vogt stated that "just as they suddenly appear, these growths may within days again disappear."

When large, they may be visible to the unaided eye. Their surface may not be smooth but rather of a granular, crumbly, uneven texture owing to accumulations of cells, and later, melanin. At times conglomerate masses may form by the coalescence of several nodules or efflorescences and may involve a large portion of the pupillary circumference (Plate XLVII, fig. 8). When in contact with the anterior lens capsule they may cause adhesions. This can be verified by

the fact that, on dilatation, pigment and exudate are found on the anterior lens capsule at their former site. In most of the present-day atlases such lesions are called "tubercles" and in subacute or chronic



FIG. 330. Efflorescences and floccules (Busacca) in iritis of unknown origin.

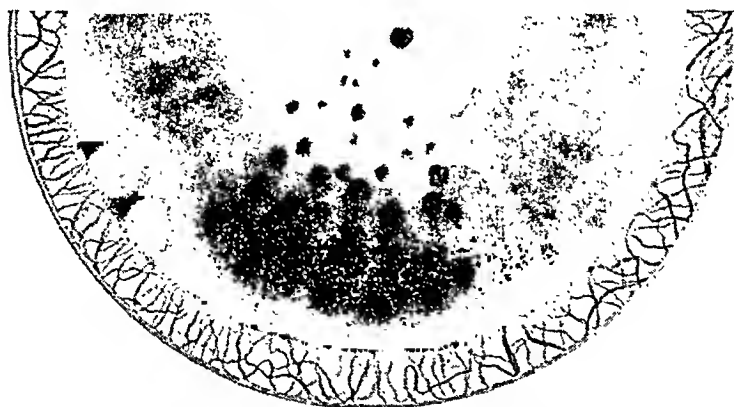
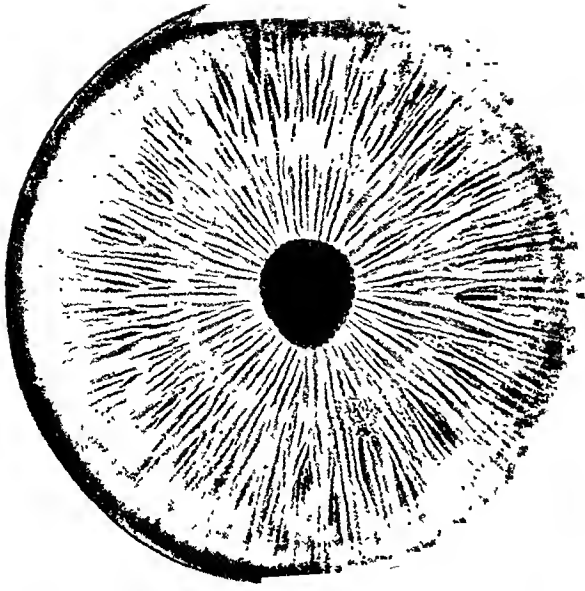


FIG. 331. Two large floccules on iris surface near the chamber angle. In the region of the posterior synechiae there was an intrastromal nodule. Large mutton-fat type of keratic precipitates. Case of Boeck's sarcoid.

cases have been considered as manifestations of tuberculosis of the iris. As previously mentioned, one must exercise caution before attributing blanket etiologic significance to them.

Mesodermic Efflorescences and Nodules. In addition to those at the ectodermic pupillary margin, efflorescences and nodules may appear not only on the stromal surface but also within the substance of the iris. As in the lesions described in the previous section, I prefer to call the smaller translucent agglomerations, "efflorescences" (those that resemble mutton-fat keratic precipitates), the slightly larger and more cystic ones, "floccules" (Busacca), and the dense, hard ones, "nodules" (Fig. 330). The first two types occur on the iris surface, like deposits, while the nodules may occur not only on the surface but also within the stroma itself (Fig. 331).

In my experience the superficially-placed efflorescences and floccules occur in the acute phases of iridocyclitis whereas the nodules tend to form in the more subacute or chronic cases. I have seen a case (Plate XLVII, fig. 5) of acute luetic iritis in which there were numerous white cystic balls (floccules), seemingly lightly attached not only to the pupillary margin but to various places on the stromal surface. Within ten days they disappeared leaving no trace (syphilids?). Vogt, who considers a large majority of the nodules to be tuberculous, described two mesodermic forms which he calls type I and type II. Type I, which he states are typical tuberculides of the iris stroma, have a homogeneous opaque appearance and may become glassy (Plate LIV, figs. 2, 3, 4). They are best seen in light colored irides where they are lardaceous and white in color, and form sharply delineated homogeneous masses, lying freely on the anterior surface of the iris. In one case they became detached and reached the lens capsule. These structures which contain no vessels are not part of the iris substance but generally lie on it. Type II is found within the iris tissue itself either diffusely or in the form of nodes (Plate LIV, figs. 1, 5). The latter thicken and become protuberant, sometimes covered by stroma. With both types, especially as the lesions age, there is a decided tendency for pigmentation to occur (Plate LIV, figs. 3, 4). Such pigment granules, either carried by the aqueous or migrating within the stroma eventually become deposited on the nodules. The strands of iris trabeculae are not visible within the nodules. If diffuse, these formations may easily be overlooked, espe-



B



C



A

FIG. 332. A. Peripheral anterior synechia resulting from slowly progressive conglomerate tubercle in iris periphery. B. Diffuse depigmentation of the pupillary area in a blue iris. (Secondary pigmentation is slight.) C. Optic section of B. Note margin of sphincter.

cially if they are situated in the vicinity of the lesser circle. In the latter case indirect illumination of the iris may reveal them as glassy thickenings within the stroma. Here, they may assume various shapes, resembling sausage-like thickenings or more often cupula-like swellings. Such masses may be extremely difficult to visualize in heavily pigmented irides. As a rule they become vascularized. Small red threadlike vessels may be seen in the neighboring stroma or running between the nodules themselves (Plate LIV, figs. 1, 5). At times these masses may become so large as to extend to the posterior corneal surface and, especially if they arise in peripheral parts of the iris, may cause varying degrees of anterior peripheral synechiae which, after organization (Fig. 332), may lead to a fibrous adhesion and obliteration of the chamber angle. Koby has noticed that in such cases, late atrophy of the iris stroma may result in a relative deepening of the anterior chamber, the free angle becoming rounded and obtuse. On the whole, nodules are less frequent in the ciliary zone than in the region of the lesser circle and pupillary area. In general, and depending on size, the intrastromal nodule leaves permanent scarring and vascularization, and is associated with atrophy of the iris. Also as mentioned before, and though difficult to detect biomicroscopically, histologic preparations have illustrated the manner in which a deeply seated nodule may extend or rupture posteriorly and cause a widespread posterior synechia.

Synechiae. The classic sign in inflammatory states of iritis is the synechia. It is interesting to note that biomicroscopy has offered little new knowledge concerning the mechanism either of posterior synechiae (adhesions between the posterior surface of the iris and the anterior lens capsule) or of the anterior peripheral synechiae (adhesions between the peripheral anterior surface of the iris and the posterior corneal surface) so thoroughly described (histologically) in the past. It would be impossible to describe all the bizarre types of synechiae. Rarely does one exactly resemble another (Plate XLVIII, figs. 1-9).

The readiness with which adhesions form between the iris and its adjacent structures has caused certain authors to liken the anterior chamber to the peritoneal cavity of the abdomen. Posterior synechiae,

PLATE XLVIII

FIG. 1. Synechiae with iris atrophy.

FIG. 2. Two solid synechiae. Lens cataractous. Pupil distorted.

FIG. 3. Atrophic iris with numerous tongue-like synechiae. Indirect retro-illumination.

FIG. 4. Toxic iritis. Smearing of the pigment at the pupillary margin. Note circulation hole at upper border of pupil.

FIG. 5. Elongated and stretched synechiae. Note opacity at point of attachment to the lens.

FIG. 6. Detached pigment particles from the seam adherent to exudative membranes which are connected to the lens.

FIG. 7. Egg-spawn conglomeration of pigment at the seam resulting from detached synechiae (luetic).

FIG. 8. Synechiae (ectropion in type). Note lens spots.

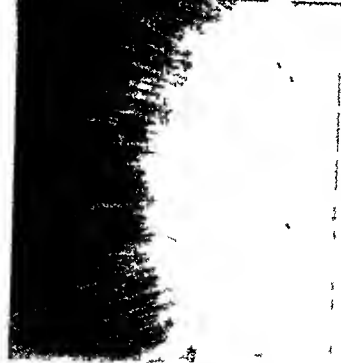
FIG. 9. Absence of the seam excepting below. Pigment and exudative membranes on the anterior lens capsule. Atrophy of iris.



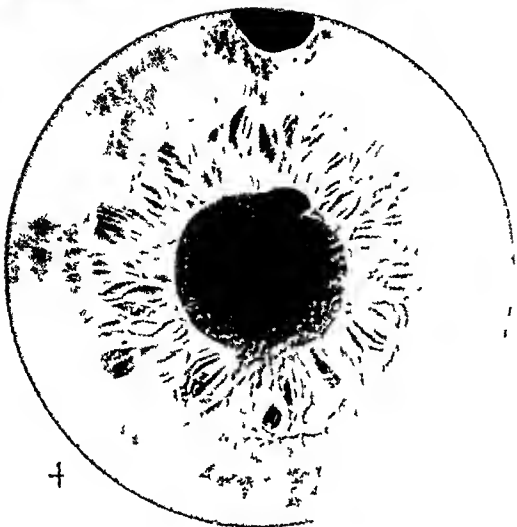
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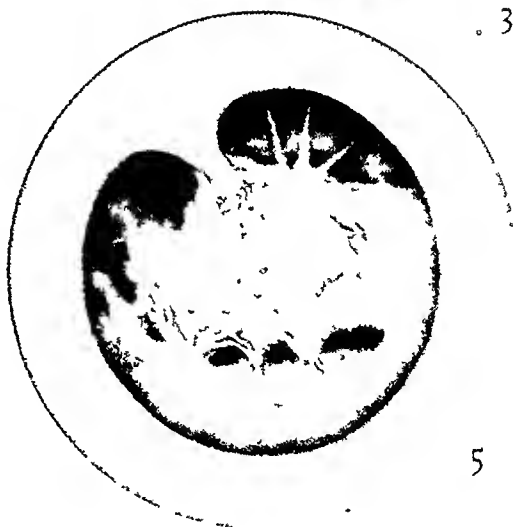
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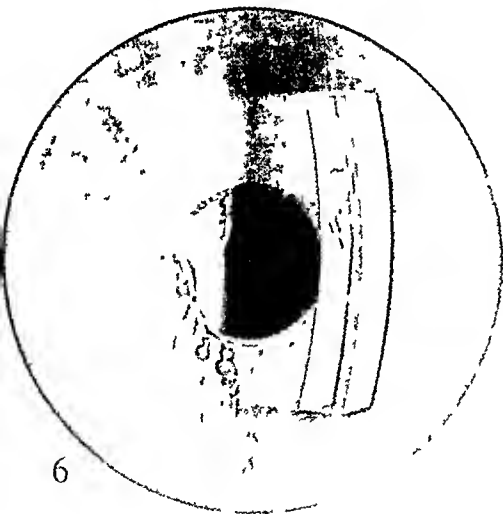
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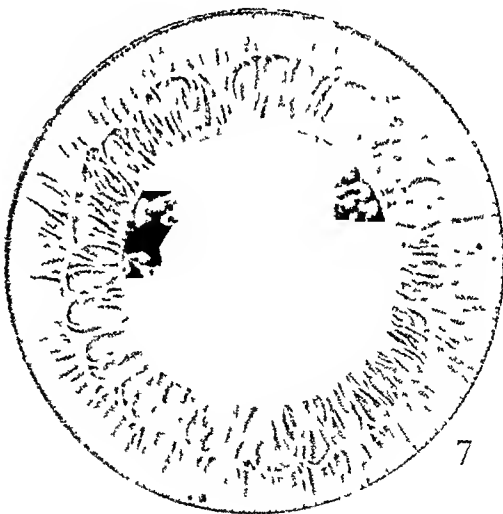
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the form commonly seen in iritis, occur at the pupillary margin where the anterior bulge of lens is in contact with the posterior surface of the iris. The tendency to miosis in iritis assists in a passive way toward adherence by affording a larger area of contact. The miosis and pupillary sluggishness, which according to my observations occasionally follows a brief period of slight initial mydriasis, is due to irritation (spasm), hyperemia, and exudation in the iris tissue. The actual adhesion, which also tends to keep the pupil small, is formed by exudate from the posterior iris surface which, on organization, produces a seal between it and the lens capsule. Direct adhesions of the iris stroma can occur to the lens capsule, in which atrophic tongue-like processes of stroma are pulled over and attached to a delicate exudative pupillary (capsular) membrane. The seam in these areas will be missing or atrophied. In a case recently observed the membrane over the lens capsule was so thin that at first it was overlooked. However, the presence of pigment granules beneath it on the anterior lens capsule, as seen by optic section, demonstrated that these granules had been deposited before the membrane had formed. How much of a role, if any, free exudate in the anterior or posterior chamber plays in the formation of posterior synechiae is not known. It is conceivable that exudate thrown out by the convection currents of the aqueous, in the narrow zone between the pupillary margin and the lens capsule, might be deposited there (similar to keratic precipitates).

It is not unusual, especially in the acute cases, for the pupil to return to its previously normal size and shape; this restoration, however, is less frequent in the more chronic granulomatous cases, particularly those characterized by the formation of nodules.

As pointed out above, the presence of an efflorescence at the pupillary pigment border does not always presage an adhesion. Those situated at the posterior (lens) side of the seam are more liable to result in synechiae, especially if several of them coalesce. Often, when the pupil is small, adhesions located here will be invisible from in front; their presence only becomes manifest after dilation (following rupture or stretching of the adhesion, either spontaneously or

artificially by the instillations of mydriatics), when they appear as traction tags or as clumps or dots of pigment adherent to the capsule (Plate XLVIII, fig. 5). With swelling of the iris, in the active stage, it is conceivable that more of the posterior surface of the iris than the pupillary margin contacts the anterior capsule. This may account for the occasional presence of radial lines of pigment dots on the capsule that can only be seen following mydriasis. After the formative periods of adhesions, attempted mydriasis may result in the appearance of a scalloped pupil, since only the part between the adhesions can dilate (Plate XLVIII, fig. 2). Also, even after organization occurs, an adhesion may be stretched, permitting moderate dilation. Frequently, after iritis has subsided and there are some remaining adhesions (posterior synechiae) the pupillary movements may not be disturbed since the stroma glides over the fixed posterior pigment layer. This can only occur provided the iris is not too atrophic and the sphincter remains active. If the use of mydriatics is instituted early, adhesions may all be separated, but as a rule they will permanently leave their telltale pigment clumps derived from the pigmented layer, attached to the anterior lens capsule. Corresponding to the size of the pupil before mydriasis and providing that more than one or two adhesions were present, an interrupted circular imprint of pigment on the capsule will leave indelible evidence of a past iritic inflammation (Plate L, fig. 1). At times, after the florid stage has subsided, fine fibrinous threads may be noticed, usually pigmented, and resembling residua of the pupillary membrane. These threads, arising either from the pupillary margin or from the pupillary region of the iris, may singly or in a branching way extend to the lens capsule or to an adjoining place in the pupillary part of the iris (Plate XLVIII, fig. 7). Others attached at one end may be found floating freely in the aqueous. Such threads are frequently seen after fetal iritis. They may be attached to flattened polar or eccentric capsular opacities. As these threads, which sometimes may be thickened or ropelike, approach the pupillary edge of the iris, they tend to broaden out. The fact that they broaden out

(leaflike) and attach to the pupillary pigment seam indicates their inflammatory origin rather than any relationship to congenital pupillary remnants. In other words they result from the stretching of posterior synechiae. With miosis, threads of this kind become kinked while after mydriasis they tend to stretch out and become taut (Plate XLVIII, fig. 1).

More extensive exudation (in the form of membranes) in this region usually forebodes broader adhesions; in the extreme state an annular synechia may seclude the pupil entirely. Patients who have not had the benefit of local treatment (mydriatics) end up with pinpoint pupils. Obviously dilation of the pupil, if kept constant (a difficult task in acute iritis), prevents the formation of the usual posterior synechiae; but at times the latter will form even when the pupil is dilated. In such cases, when there is associated atrophy of the tissue, the pupil remains wide permanently and loses its reactions (Plate XLVIII, fig. 8).

Traction on the excrescences, themselves, tends to pull them off the iris attachment or to cause their disintegration with the resulting migration of pigment. The synechia itself tends to become covered with this pigment dust so that it soon becomes impossible to recognize any of its structural makeup. However, all the scar tissue of the adhesions does not necessarily become pigmented. Often a structureless whitish band (broad synechia) or node of organized tissue may be seen attached to the capsule (Plate XLIX, figs. 1, 4). These bands may run circumferential to the pupillary edge for longer or shorter distances immobilizing the pupil in part or throughout the extent of the band. The pupillary edge in the region of such bands reveals the absence of normal excrescences, atrophy of the stroma and fine deposits of pigment grains. At times one or more of the excrescences may be attached to these membranes. The attachment of the membranes to the lens is not always smooth but pleat-like so that the filmlike membrane over the capsule is thrown into delicate folds (seen best in the shagreen area, zone of specular reflection). In other cases the membrane may detach itself from either the lens or iris in such a way that the free end curls over to its place

of attachment. The formation of delicate meshworks of fibrinous exudate on the anterior lens capsule probably results from contraction and organization of exudate. Frequently, thickenings at the places of crossings of fibers forming the net results in starlike structures in which pigment granules may or may not be incorporated (Plate LXXIII, figs. 1-6). Also it is not unusual to see small whitish deposits (threads or stars) on the anterior lens capsule in the vicinity of an adhesion, or even when no synechiae are present.

Migration of pigment may extend into the sphincter zone of the stroma or may be carried to the boundaries of the anterior chamber by the aqueous currents. At the site of the adhesion, especially when it is small, the excrescences are missing, and a slight degree of entropion will be recognized. Annular synechia is generally the result of attacks of plastic iritis. Biomicroscopic inspection will practically always reveal a membranous exudate extending from the iris to the lens capsule (Plate LI, figs. 1, 7, 8). Such extensive membrane formation results in occlusion of the pupil with or without seclusion; the membrane frequently becomes vascularized. In ordinary isolated synechiae there is little tendency for vascularization to occur at the point of adhesion although it may be present in the adjacent stroma.

In *toxic iritis* a smearing of pigment between the pupillary border and the lens capsule may occur (a form of posterior synechiae) (Plate XLVIII, fig. 4). I have often seen such deposition of pigment at the pupillary border in the iritis that not infrequently follows trephine operations for the relief of chronic simple glaucoma; central vein thrombosis; after retinal detachment; and in diabetes. In retinal separations, iritis and complicated cataract as well may result from the toxic effect of the subretinal fluid which seeps into the preretinal region. This type of "pigment-smeared" adhesion is not as apt to be a localized synechia through the medium of an efflorescence as is seen in "infective" iridocyclitis, but is more diffuse. It is possible, according to Vogt, that toxic substances in the aqueous cause suspension of pigment which is in turn deposited. Characteristically, in cases of toxic iritis there is an absence of keratic precipitates. But with secondary infection corneal precipitates are found. This form of

smear pigment adhesion may be annular (*seclusio pupillae*), but usually atropinization demonstrates one free area, generally above, in the form of a circulation hole, often resembling a horseshoe. Such an opening may serve as an adequate communicating channel between the anterior and posterior chamber. A gap (circulation hole) in an almost complete annular synechia also occurs in chronic types of exudative iridocyclitis (page 897), especially those of tuberculous origin. In these cases smearing of pigment at the pupillary margin (as in toxic iritis) is not found, but thin exudative membranes are seen extending from the pupillary margin over the lens capsule. Vogt has compared the mechanism of "circulation hole" formation to that which occurs in a running stream which does not freeze. In other words it is difficult for exudate to seal off the current in a region where there is active circulation of fluid, especially as the "secluding" mechanism is gradual. The association of toxic iritis and hypotony is interesting.

Nodular formations in the peripheral parts of the iris or exudative fibrinous masses may undergo organization and form anterior peripheral synechia (Fig. 332 B). These obliterate the chamber angle not only by their volume and mass but also by causing adhesions between the iris stroma and posterior corneal surface.

PIGMENTARY CHANGES

As in senility, congenital alterations, or with atrophic disorders, inflammatory states of the iris are characterized by disintegration of its pigment (Plates XLIII, fig. 1; XLVIII, figs. 1, 3). This is recognized by depigmentation, scattering, and migration not only of the retinal pigment but also of the melanophoric stromal pigment (Fig. 315).

The degree of pigmentary changes (both retinal and stromal) depends on the intensity of involvement, chronicity, local resistance, the original iris coloration, and possibly on the age of the patient. Edematous swelling of the pigment-bearing cells results in their disruption, which is followed by scattering of the contained pigment. Possibly some of the free pigment is phagocytosed by remaining

cells. The tendency of pigment to proliferate and the property of these cells to act as histiocytes is well known. A combination of some or all of these factors may serve to explain the pigmentary changes seen in iritis, which if marked cause the iris coloration to change. As a rule in inflammatory lesions the posterior retinal pigment layer is first affected. Depigmentation of the stroma usually accompanies atrophy of the mesodermal iris and may be a later or concomitant manifestation (Plate XLVIII, fig. 9).

Degeneration and depigmentation of the retinal iris layer affects not only the seam of the pupillary border but also, either in part or almost completely, the posterior pigmented iris surface. Examination of the pupillary border in some cases may reveal areas in which the excrescences are missing* and other areas of depigmentation not unlike the ordinary senile marginal atrophy. The latter is best seen by retro-illumination, the affected pupillary border being observed by light reflected from the lens as the focused beam passes into the pupil. In addition to atrophy in the regions of posterior synechiae, a secondary increase of pigment may be found. As in glaucoma, attraction of pigment to the pupillary border area (secondary pigmentation) may also occur in chronic cases of iridocyclitis (Plate LI, fig. 6). In these, migration of retinal pigment to the pupillary zone of the iris may be marked. Pigment destruction of the posterior iris layers results in a typical moth-eaten appearance when viewed by either retro-illumination or by transscleral illumination. These may be confined to local areas or may become so extreme as to leave only a shell of iris substance behind (Plate L, figs. 4, 8, 9). As mentioned before, except in special cases, in which one can see into the bottom of a deep crypt, it is impossible biomicroscopically to see the posterior pigmented iris layer. When this layer is intact, it does not allow any of the reflected light from the lens to pass back through the iris structure to the eye of the observer. When there is any destruction of the posterior pigment layer, passage of the focal beam into the lens via the pupil allows the reflected light to pass back through the iris at these points (retro-illumination) (Plate L, fig. 1). This

* The deposition of pigment clumps on the anterior capsule following synechiae is discussed on page 846.

affords a sort of negative image of the lesion (shadow effect) in which lighter translucent areas appear in the otherwise darkened tissue. Such transilluminable areas may be in the form of punctate spots, striae, or may involve a sector of the iris, or the whole iris diffusely.

Owing to the fact that the anterior surface layer may apparently be only slightly involved,* diffuse or direct focal illumination of the iris may fail to reveal the amount of atrophy of the posterior pigment layer; thus, unless retro-illumination is used even extensive areas of atrophy may be overlooked. These "holes" of the iris were first described by Oblath (1899) who saw them following iritis in syphilis and variola.

As a result of disintegration, iris pigment granules may be found deposited not only on and in its stroma but also transported by the aqueous to the posterior corneal surface, the chamber angle, and the lens capsule. This occurs spontaneously following iritis and may follow trauma or surgery also. Small residuals of exudates on the lens capsule in the form of threads or stars also attract pigment grains, which change the original whitish appearance. In some cases it appears as if the iris is covered by fine brown or black spots resembling soot. This type of pigmentation is usually most marked in the pupillary zone, but the ciliary zone may show it as well. Extensive superficial iridic deposition of this type may result in a form of heterochromia. A blue iris may assume a dirty yellowish greenish or light brown color while a light brown iris may become darker. One can only speak of heterochromia in these cases provided the iris of the fellow eye is normal.

Localized areas of vitiligo may follow iritis. This results from atrophy of the superficial layers of stroma, especially of the anterior border layer, which in certain cases may entirely disappear. It is most

* As mentioned in the paragraph on atrophy (page 857), the formation of such "holes" in the posterior pigment layer must depend to a degree on the stromal affection. It is conceivable that if the deeper leaf of the stroma is affected chiefly, the slighter involvement of the overlying surface tissue may obscure it, to the end that by diffuse or direct focal illumination this part of stroma obscures the deeper destruction. However, indirect illumination will usually reveal a discolored area of stroma overlying these holes or areas of atrophy of the posterior pigment layer.

striking when it occurs in colored irides. Here a whitish area of deeper stroma may be seen, usually sprinkled with pigment granules (Fig. 332 B; Plate LI, fig. 6). At times the pigment from such areas migrates to and is heaped near the pupillary area. However, localized vitiliginous spots or areas are a more frequent occurrence in glaucoma, especially after acute attacks. I can recall many cases in which this occurred to marked degree following antiglaucoma surgery (Fig. 323). Also it is often noted after simple iridectomy in the region of the pillars. Incarceration of the iris pillars predisposes them to vitiliginous changes. High intra-ocular pressure probably results in pressure necrosis of the tissue. Vogt has shown that such increases of pressure also affect the lens in that damage to the capsular epithelium may be the forerunner of glaucomatous cataract. A varying degree of depigmentation of the iris is also common following trauma, particularly after perforating injuries.

The occurrence of disintegrated iris pigment in the angle of the anterior chamber has been known histologically for a long time and recently has been verified gonioscopically. However, the importance of this finding in regard to the etiology of glaucoma (primary or secondary) is still undecided (see Vol. I).

Disintegration of red blood cells after trauma or hemorrhagic iritis may also result in dispersal of hematin and changes in iris coloration (see Herpes, page 871).

CHANGES CONSEQUENT TO IRITIS AND IRIDOCYCLITIS

It happens not infrequently that after subsidence of an attack of iritis or iridocyclitis no macroscopically visible changes remain; however, it is very unusual not to find some residual microscopic alterations. The importance of biomicroscopic examination for this purpose cannot be overemphasized. The presence of pigment deposits on the anterior lens capsule, especially those near the pupillary margin, is very significant. These deposits usually have the form of rough clumps, but occasionally dotlike pigment grains may be seen arranged irregularly or in the form of stars, rings, or radial lines (Plate LXXIII, figs. 1-6). Dilation of the pupil is frequently necessary to

disclose the finer ones, which may derive from the ridges of the posterior iris surface, and may be bound to the lens capsule by delicate exudates. Close observation of the pupillary pigment seam may disclose atrophy of the border or absence of an excrescence or two in the neighborhood. Exudate deposited on the anterior lens capsule, especially near the pupillary margin, may after a time become transformed into fine threads (by contraction) or starlike figures. When delicate, these may only be seen biomicroscopically.

More rarely, a few fine pigment grains (keratic precipitates) or dots of fibrin may be found on the posterior corneal surface long after the inflammation has subsided. However, in recurrent iritis with marked exudation or in long standing chronic cases (granulomatous), keratic deposits may remain permanently.

Ordinarily the tendency toward the absorption of exudative keratic precipitates is great but, as previously mentioned,* older residua (especially in iridocyclitis) become crenated and pigmented. Also the larger mutton-fat-like and plastic exudates tend to leave areas of increased relucency not only on the posterior corneal surface but in the deeper layers of the corneal stroma itself. A notable example of this is seen in the iridocyclitis associated with interstitial keratitis. (See Vol. I, page 500). Examination of the posterior lens capsule and vitreous may also reveal precipitates and pigment even in the so-called "light" cases (Fig. 326). It goes without saying that the presence of posterior synechiae, which may be seen grossly with varying degrees of immobilization of the pupil, is immediately diagnostic of a past iritis. However, severe single or recurrent attacks of iritis as well as the low-grade chronic forms are generally characterized by changes within the iris tissue itself. From the standpoint of biomicroscopy they result in (1) pigmentary changes and (2) atrophy of the iris stroma itself.

IRIS (STROMAL) ATROPHY (POSTINFLAMMATORY)

Atrophy of the iris stroma not only occurs as a sequel to severe recurrent or chronic inflammations (iritis) but also follows trophic

* See Volume I.

PLATE XLIX

FIG. 1. Localized area of iris atrophy. Broad adhesion with exudate. Viewed by retro-illumination and direct focal illumination.

FIG. 2. Radial areas of iris atrophy—high-power view.

FIG. 3. High-power view of the lesion seen in Figure 1. Diffuse illumination.

FIG. 4. Details of broad adhesion seen in Figure 1.

FIG. 5. Iris atrophy of the pupillary margin and stroma. Retro-illumination.

FIG. 6. Atrophy of the pupillary border and stroma. Rarefaction and condensation of the stroma. Posterior pigment layer showing through. Diffuse illumination; high power.

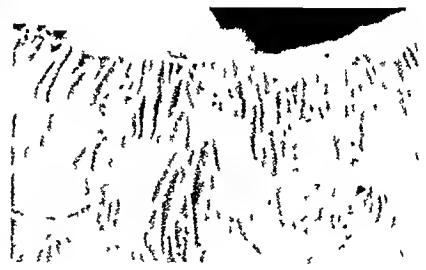
FIG. 7. Chronic iritis. Iris atrophy with vessels migrating over the anterior lens capsule. Retro-illumination.



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disturbances, trauma, and increased intra-ocular pressure. Senile atrophy, which in a sense may be considered as physiologic, is discussed on page 771. In general, the changes which produce this picture of iris atrophy are due first to pigment degeneration (and the secondary dispersal of pigment granules) and second to alterations within the stroma itself. The pigmentary changes which are discussed on page 849 involve not only the posterior ectodermal iris layer but also, especially in the colored iris, the stromal pigment. Depigmentation of the posterior iris layer results in "holes" or areas of lesser optical density illustrated by transpupillary retro-illumination (Plates XLIX, figs. 1, 7; L, figs. 1, 3, 6, 9). This occurs not only at the pupillary margin but also in a regular or irregular fashion, either sectorially or completely, anywhere on the entire extent of the posterior iris surface. Examination by direct focal light may ordinarily fail to reveal atrophy of the posterior pigmented iris layer; the overlying stromal layer, which may only be partially affected, diffuses the light and prevents observation of the holes. Considering the fact that the posterior pigmented layer receives its nutrition from the stromal vessels, it would seem that interference with these vessels, either primarily or as a result of stromal involvement, would of necessity cause changes in the underlying pigment layer. This is reminiscent of a similar relationship between the choroid and retina; the layer of pigmented epithelium and outer retinal layers are nourished by the choroidal vessels and hence react secondarily to affections of the choroidal circulation. The formation of these areas of posterior pigment degeneration (holes) after iritis depends not only on interference with the circulation, but mechanical factors also play a part because fixation of the iris by synechiae favors atrophy by traction. Undoubtedly, localized areas of atrophy involving the stroma and the posterior pigment layer as well can follow resolution of nodules (especially tubercles) and papules. Indirect illumination at times will better demonstrate the stromal changes over such a depigmented area than direct focal illumination.

Degenerated pigment granules may migrate and may be deposited within or on the iris stroma, or they may be transported by aqueous

PLATE L

FIG. 1. Iris atrophy. Surgical coloboma. Pigment on the anterior lens capsule. Anterior complicated cataract. Retro-illumination.

FIG. 2. Detailed view of changes following iritis. Temporally, the iris stroma is condensed and flattened. New vessels. Crypts obliterated.

FIG. 3. Brucellosis. Areas of iris atrophy.

FIG. 4. Marked atrophy of the iris—complicated cataract.

FIG. 5. Heterochromia iridis syndrome of Fuchs. Keratic precipitates. Aqueous flare. Complicated cataract. Atrophy and discoloration of the iris.

FIG. 6. Vitiligo of the iris—Herpes.

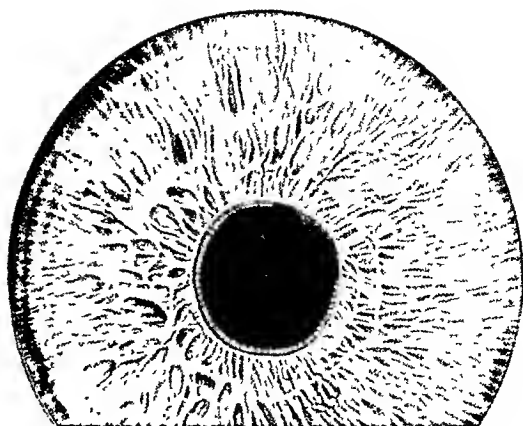
FIG. 7. Localized area of atrophy with marked loss of iris substance following iritis.

FIG. 8. Diffuse view of iris following sympathetic ophthalmia.

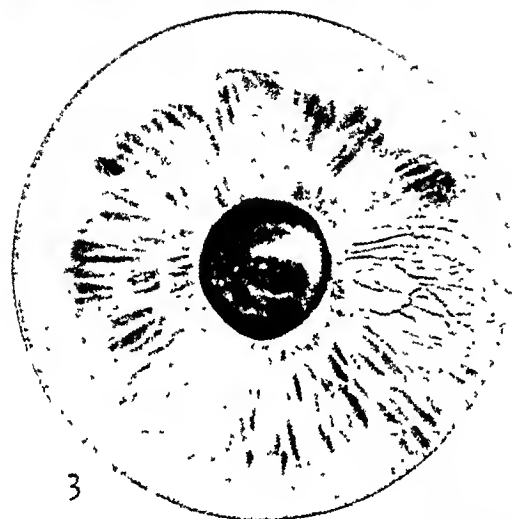
FIG. 9. Same case as Figure 8. Viewed by retro-illumination. Pupil secluded. Vessels extending over the anterior lens capsule. Complicated cataract.



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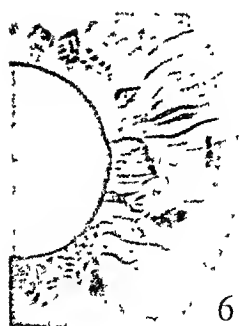
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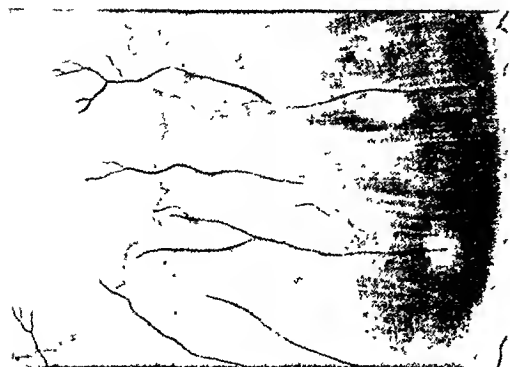
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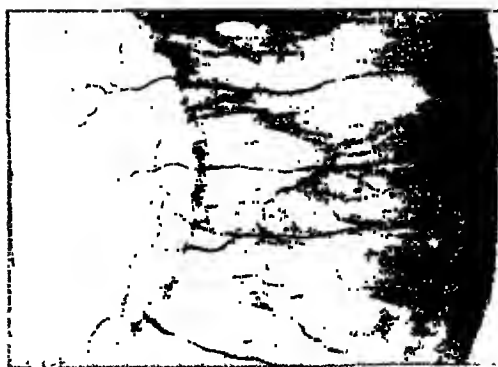
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either out of the globe or to any portion of the chamber boundaries (Plate XLIX, fig. 6). Depigmentation of the stromal melanophores is best seen, as would be expected, in brown irides. In the latter localized vitiliginous spots or a generalized depigmentation of the surface stroma may be found involving a sector of the iris or its whole surface (Fig. 332 B, C). Depending on the extent and degree of depigmentation, color changes result. In some dark brown irides, vitiliginous areas appear dirty white and structureless; but usually these areas are sprinkled with fine pigment granules (Plate LI, fig. 8). While in others, in which the anterior leaf is chiefly involved, the exposed depigmented areas reveal whitish or bluish radiating trabeculae (Plate L, figs. 6, 7). Deeper destruction may expose the posterior iris pigment layer covered by a few completely or partially depigmented stromal fibers. At times a few free trabecular fibers may float loosely in front of the atrophic area or they may form an irregular mat. As a rule, in postinflammatory iridic stromal atrophy only certain sectors in an individual iris will reveal extensive destruction of tissue (i.e., where the lesion had been most intense), the neighboring areas showing relatively fewer degrees of atrophy (Plate L, figs. 6, 7). An atrophic sector of the stroma may extend from the periphery to the pupillary margin (including the seam) or only the pupillary or ciliary part themselves may be involved.

In atrophy of the pupillary zone, owing to the thinness of the stroma, it is frequently possible to see the brown areas of the posterior iris layer (Plate XLIX, fig. 6). This thinning and rarefaction in the pupillary zone may become so extensive as to permit an uninterrupted view of the posterior pigmented layer, a condition which may be mistaken for ectropion. However, a few stromal strands and the irregular line of the lesser circle usually remain. Vogt has termed this change, resulting from inflammatory atrophy of the stroma, as "circumpupillary exposure of the retinal iris layer." In other cases the atrophic pupillary zone, sometimes the whole iris surface, may be sprinkled with fine black spots of pigment resembling soot. These dark grains may also be deposited on the posterior corneal surface. Likewise, pigment which is freed in a vitil-

iginous process of the iris (especially after acute attacks of glaucoma) may by their weight be deposited in the bottom of the anterior chamber similar to a hypopyon. Vogt called this melanohypostasis.

In lighter colored irides the depigmented area may appear greenish, yellowish or whitish. These types of stromal depigmentation are also seen in glaucoma and after virus infections, e.g., herpes, variola, certain exanthems and the like. The close association between pigment and nervous tissue is of interest in this connection. Atrophy of the iris stroma itself produces a variety of clinical pictures depending not only on the type or intensity of the involvement but also on the original coloration and structure. In dark brown irides with little visible trabecular structure, modification of the surface markings and thinning and flattening may be the only changes noticeable. More extensive damage results in stromal depigmentation and the appearance of vitiliginous areas. Vitiliginous spots may appear in the stromal periphery with no apparent alteration or destruction of the pupillary pigment.

Stromal atrophy is best exemplified in blue or light colored irides, especially if the pupil is not dilated. The iris appears thin and the surface flattened and condensed to one plane, so that the normal difference in thickness between the pupillary and ciliary portions no longer is apparent (Plate XLIX, fig. 3). The trabeculae are stretched out and pulled in a radial direction. The line of the lesser circle (frill) which usually runs concentrically to the pupil tends to become angularly elongated and to be retracted peripherally* (see Fig. 332 B and C). At various places between the stretched fibers, reddish threadlike vessels will be seen. The fibers lose their fluffy appearance and appear frosted. The crypts become less prominent or are obliterated (Plate L, fig. 2). The atrophic trabecular fibers appear whiter, especially in the ciliary zone where the condensation of the anterior and posterior leaf will prevent the diffraction phe-

* This picture is contrasted to a type of iris atrophy seen in absolute glaucoma in which atrophic changes in the pupillary zone result in a narrowing of this zone (dilation of the pupil). Here a widening (ectropion) of the pupillary seam is seen, and the frill is drawn closer to the pupillary margin. Atrophy (necrosis ?) of the ciliary zone of the iris may be so marked as to expose the white sclerotic arterial walls and also dilated and tortuous red veins (glaucomatous rubeosis).

nomenon that ordinarily gives the iris its bluish tinge. However, because of its thinness the pupillary area (its stroma substance chiefly consisting of the posterior leaf) may appear darker and more bluish. Localized areas of atrophy, such as result, for example, from the resolution of nodules, may leave darker discolored spots; but with the optic section it will be seen that the stromal thickness is thinned out, leaving only a narrow padding of tissue over the posterior pigmented layer (Plate XLIX, fig. 7). In other words, owing to actual loss of stromal substance, an excavation occurs.

Immobilization of the sphincter by adhesions or by the organization of exudates, leads to its atrophy or disappearance, so that proximal illumination no longer brings out its ordinarily sharp peripheral border. Also, the pupillary edge, which may be sharp or fringed, can be drawn backward (i.e., towards the lens) together with the sphincter; this results in a slight degree of entropion (Plate XLIX, fig. 3).

In cases of postinflammatory iris atrophy of long standing, ectropion of the posterior pigment layer can occur (Plate XLVIII, fig. 8). Evidently atrophy of the stroma, combined with traction of organized membranes over the iris surface, serves to evert and draw the pigment over the iris.

An interesting form of rapid tissue dissolution occurs in necrosis of iris (Samuels).⁵⁹³ This is the sequel to a sudden cutting off of the blood supply, e.g., by trauma, glaucoma, localized thrombosis, or tumors, or by an overwhelming toxemia generated in an acute or massive bacterial infection, especially following perforating wounds or in association with necrotic tumors. Histologically, necrosis differs from atrophy in that in the former, acute swelling with destruction of nuclei results in a ghostlike appearance of the tissue while in the latter, although the tissue becomes thin and condensed, it still retains some of its cellular aspects. Biomicroscopically, it may be impossible to differentiate between these two processes. Depending on the circumstances, one could surmise that many cases that are diagnosed as iris atrophy might have been the result of necrosis. In those cases in which so much of the iris tissue is destroyed that only a trans-

PLATE LI

FIG. 1. Diabetic rubeosis of the iris. Secondary glaucoma. Complicated cataract.

FIG. 2. Rubeosis iridis—glaucoma.

FIG. 3. Rubeosis iridis—chronic iritis.

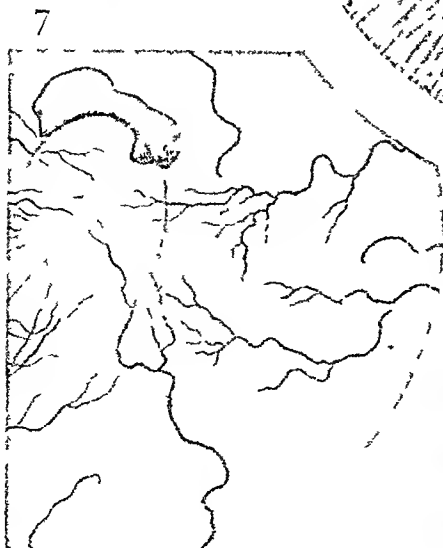
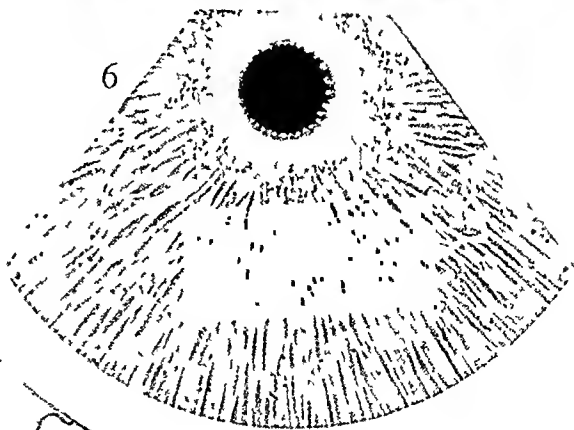
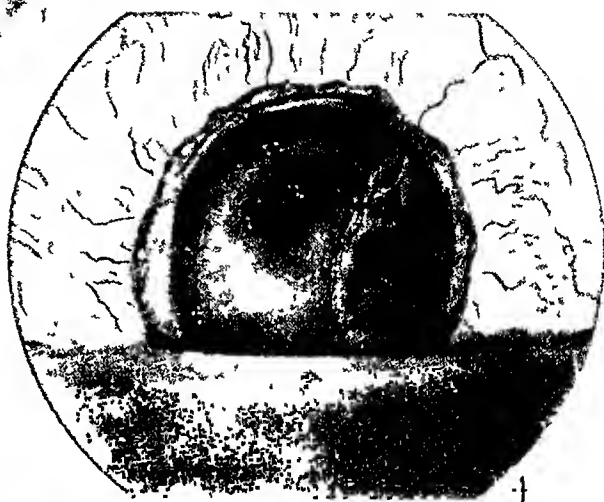
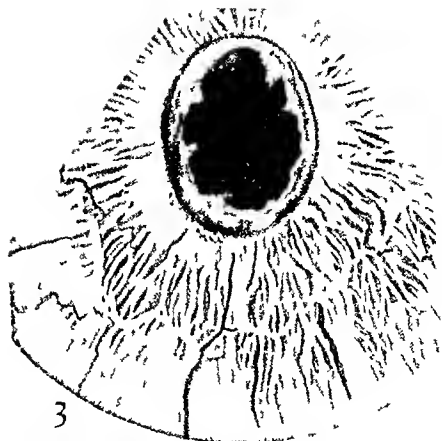
FIG. 4. Rubeosis iridis glaucomatosa. Hyphemia. Following thrombosis of the central retinal vein.

FIG. 5. Localized iris atrophy following glaucoma.

FIG. 6. Secondary pigmentation of the iris (glaucoma).

FIG. 7. Iris atrophy—rubeosis iridis. Secondary glaucoma. Vascularized exudative membrane over the lens capsule.

FIG. 8. Same case as Figure 7. The iris is viewed by optic section. Observe vaulting of iris below.



parent veil remains (retro-illumination), one could perhaps be justified in speaking of iris necrosis rather than atrophy.

In postinflammatory as well as in other types of atrophy of the iris, visible vessels are present (Plates XLIX, figs. 3, 7; LI, figs. 1, 2, 3, 4, 7, 8). For the most part these appear to be the ordinary trabecular vessels which have become permanently engorged. They have as a rule the characteristic radial direction, although as they approach the lesser circle, branchings may occur; then, meeting other divisions, the vessels may follow the direction of the lesser circle (Plate LI, fig. 2). From there more or less radial branches may extend to the sphincter area. With more extensive stromal atrophy new-formed vessels become manifest. They tend to be larger than the trabecular ones, have no sheathings (uncovered) and run irregularly not only within the stromal substance but also on the iris surface. In addition, they frequently appear as convoluted tubules or even varices. With recurrence of an attack of iritis or even spontaneously, rupture of these vessels results in hyphema with its attendant complications (page 871).

In 1883 Fuchs⁴³⁵ demonstrated histologically that new-formed surface vessels may lie in an exudative membrane that covers the iris surface and frequently extends over the pupillary area (Plates XLIX, fig. 7; LI, figs. 7, 8). These vessels, instead of terminating or looping back at the pupillary margin, continue over it in the membrane which partly or completely covers the pupil. An organized exudative membrane of this kind, which extends over the pupillary margin, can result in seclusion; if it covers the pupil entirely, it may cause occlusion (Plate LI, fig. 7). Unless a "circulation hole" remains, this will be followed by iris bombé and secondary glaucoma. Such pupillary membranes are commonly found after severe or chronic involvements, especially with the granulomatous infections, and may not only cover the surface of the iris wholly or in part but may also form on the posterior surface of the iris, causing complete or extensive adhesions. The presence of a surface membrane contributes to the discoloration of the iris (Plate LI, fig. 8). Ectropion of the posterior retinal pigment layer can occur as a consequence

of long standing iritis, especially in degenerated eyes after prolonged periods of increases of intra-ocular pressure. This sequel of events occurs frequently following absolute glaucoma, in cases of detachment of the retina, intra-ocular hemorrhages and tumors as well as from long-standing inflammations. Sclerosis of iris fibers together with traction of organizing exudate may be the causal factors in producing the eversion of the posterior iris pigment layer (see congenital ectropion, page 789). The degree of ectropion varies from case to case; collar-like, sectorial or apron-like forms may occur. The peripheral limits of the ectropion on the iris surface may be irregular, resulting in a zigzag dentate edge (Plate LI, fig. 4).

As a terminal picture, seclusion of the pupil may lead to "vaulting" or folding of the iris (Plate LI, fig. 8). The aqueous trapped in the posterior chamber pushes the iris forward toward the cornea until the anterior chamber is practically abolished, except for a small area in the vicinity of the pupil. Extensive adhesions (synchiae) in sectors in which the whole posterior iris surface is bound down, i.e., from the pupillary area to the root of the iris, may prevent the iris from being pushed forward uniformly; thus the iris has a folded appearance. This naturally would prevent the formation of typical iris bombé.

Chapter Twenty-One

SPECIFIC TYPES OF IRITIS

THE protean biomicroscopic aspects exhibited by an endogenous iritis or iridocyclitis, even those consequent to the same cause, in itself should caution one against presuming an etiologic diagnosis on the basis of the ocular picture alone. For example, in gonorrhea, syphilis, or tuberculosis forms may occur which range from the so-called abortive type to simple, exudative, plastic, or proliferative types. Although clinically many of the so-called rheumatic iritides (e.g., those resulting from focal infection, possibly depending on the presence of local tissue allergy), the gonorrheal, and luetic types of iritis tend to run an acute course, while others (e.g., the tuberculous type) are characterized by chronicity; but actual statistics assembled by numerous workers indicate a wide variability in the intensity of involvement. This is not unexpected when the pathology of the lesions is considered. Therefore, the clinical or biomicroscopic appearance must never be taken by itself as absolutely pathognomonic. In my experience, thorough medical survey all too frequently fails to reveal the source or nature of the infection in a considerable proportion of endogenous cases. In these we are forced to rely on any suggestive aid afforded by the clinical appearance. It is here that the biomicroscope may prove of incalculable value as will become evident from the consideration of some variations occurring in special types of iritis. For the purposes of orientation I include a classification (modified after Duke-Elder) of the various forms of exudative iritis, but I shall describe only those that are of special interest from the standpoint of biomicroscopy.

- I. Specific infections
 - A. Herpes
 - B. Gonorrhea
 - C. Syphilis
 - D. Tuberculosis
 - E. Brucellosis
 - F. Leprosy
 - G. Trypanosomiasis
- II. Syndromes of doubtful etiology
 - A. Sympathetic ophthalmitis
 - B. Uveoparotitis
 - C. Boeck's sarcoid
 - D. Heterochromic iridocyclitis
 - E. Uveitis with alopecia, vitiligo, poliosis, and dysacusia (Vogt)
- III. Fungus infection (mycoses)
 - A. Aspergillosis
 - B. Blastomycosis
- IV. Iritis and iridocyclitis due to irritants
 - A. Physical agents; chemical or burns
 - B. Nodosa

HERPETIC IRITIS

In most cases, whether due to herpes simplex or herpes zoster, the iritic irritation is attendant on a corneal or conjunctival lesion, i.e., a reaction secondary to the point of inoculation of the virus. This may cause an ordinary simple or exudative type of iritis having no distinctive features—in other words, the diagnosis will depend on the presence of conjunctival or corneal involvement with or without the associated skin lesions (zoster). Or, more rarely, specific eruptive lesions on the iris may occur. It is my impression that the latter is more common with herpes zoster than with herpes simplex. Such an iritic lesion, corresponding to that of the skin, appears as a small condensed area, slightly discolored, swollen, and surrounded by congeries of dilated veins. In a recent case, in which there were

several lesions on the skin of the forehead above the involved highly inflamed, painful eye, there was a small lesion at the temporal corneal limbus. Extending from the lesser circle of the iris to the pupillary margin there was a small highly vascularized raised area. At the adjacent pupillary margin the iris margin was bound down to the lens capsule by a broad pigmented adhesion. The aqueous flare was marked, and there was a considerable number of keratic precipitates. After about six weeks the acute symptoms abated somewhat, but the eye remained congested and photophobic for about three months. It was not possible to separate the adhesions by mydriatics. Six months after onset the eye became entirely quiet, leaving not only a peripheral corneal scar involving the anterior corneal stroma but also a punched out circumscribed depigmented area in the iris with several threadlike vessels remaining in the neighborhood (Plate LII, fig. 1). The characteristic vascular reaction in the iris, when it is intense, may lead to a hyphemia which is slow in absorbing (Plate LII, fig. 2). Lowenstein⁵²⁷ described a case of this type occurring in a 33-year-old man who had five attacks within four years. Of these, four occurred in the right eye and one in the left eye. This case was characterized by violent, recurring attacks of a papular iritis associated with severe neuralgic pains limited to the affected side, and by intense ciliary injection; this was followed by anterior chamber hemorrhage each time. The pain always abated with the appearance of the hemorrhage. Examination of the right eye at the time of the last attack revealed an edematous cornea with two paracentral maculae. Three-fourths of the anterior chamber was filled with blood. After a paracentesis it was possible to obtain a better view of the iris. There were numerous swollen papular areas, in both the pupillary and ciliary zones, distributed irregularly over the surface of the greenish yellow iris. These swollen areas were covered with new-formed vessels, appearing as bright red spots. There were small stromal hemorrhages about these clusters of vessels. The pupil showed many synechiae, with pigment deposited on the anterior lens capsule. The iris as well as the posterior corneal surface were likewise

PLATE LII

FIG. 1. Herpetic iritis. Optic section through localized lesion, showing loss of substance.

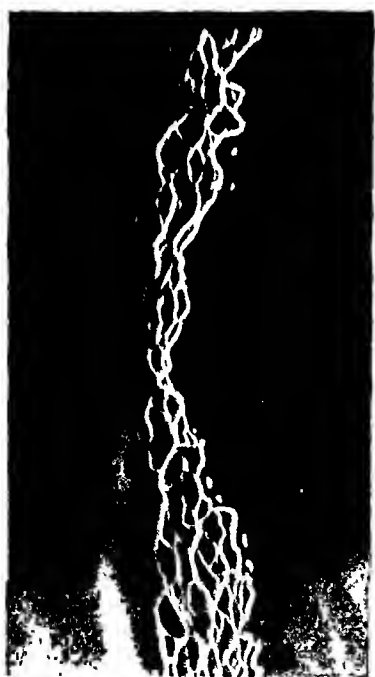
FIG. 2. Herpetic iritis with recurrent hyphemia. Iridectomy. Complicated cataract. (After Gilbert.)

FIG. 3. Chronic plastic iritis. Small gonococcal. (After Kruckmann.)

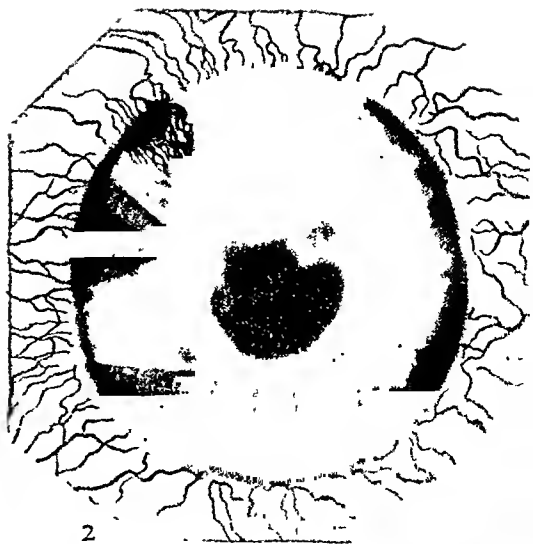
FIG. 4. Recurrent iritis. Old small adhesion nasally above, fresh exudate below. Iris atrophy (*gonococcal*).

FIG. 5. Roseolae of the iris (*lues*).

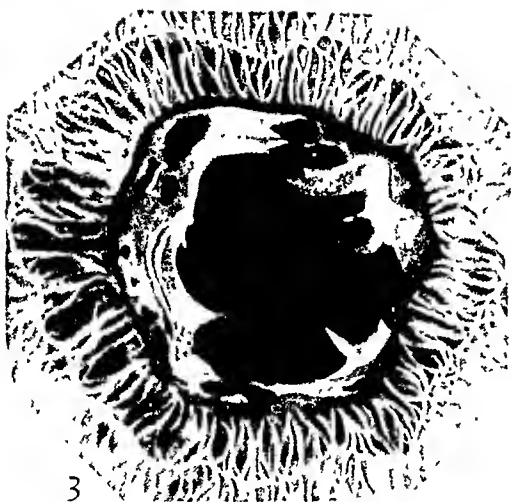
FIG. 6. Papular iritis (*lues*).



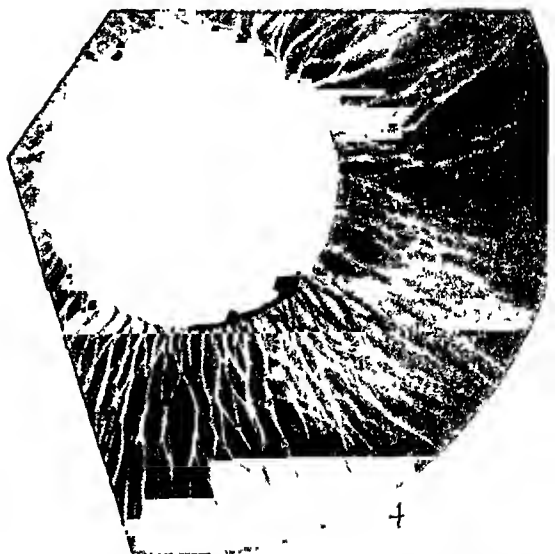
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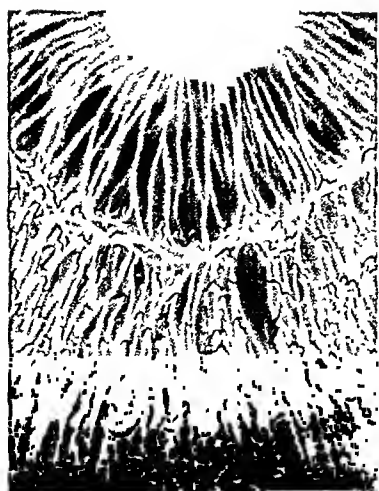
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3



4



5



6

sprinkled with pigment. A dark hyphemia occupied the lower part of the anterior chamber.

In summarizing the findings in herpetic iritis, Gilbert⁴⁵⁰ mentions: (1) severe neuralgic pains, (2) circumscribed swelling of the iris corresponding to the eruptions of the skin, (3) general or localized hyperemia of the iris, especially within the lesser circle, and (4) single or recurrent hemorrhages in the anterior chamber, after which the pain decreases. In severe cases, with recurrent hemorrhages, blindness results after seclusion or occlusion of the pupil by the establishment of absolute glaucoma. According to Meller (1920) hemorrhages may be marked by a hypopyon. In milder cases the hemorrhages are absorbed quickly and resolution with healing follows.

GONOCOCCAL IRITIS AND IRIDOCYCLITIS

The exudative form of gonococcal iritis is of special interest to the biomicroscopist because of its characteristic appearance. According to Byers³⁴ this form occurs in about 10 per cent of cases of gonorrheal iritis. The remainder are more or less equally divided between the simple and plastic forms* (Plate LII, fig. 3).

The acute exudative type is marked by a profuse elaboration of a jellylike fibrinous mass which at times may fill the entire anterior chamber and simulate, on hasty examination, an anteriorly dislocated lens (Fig. 324). Such a translucent gelatinous formation usually hides the pupil when first seen. Dilation, however, rapidly changes the picture, in that within the following few hours the exudate drops down to the bottom of the anterior chamber and the pupillary margins become visible. Evidently the exudate in the beginning is attached to the pupillary border. In other cases the mass

* Personally I have not had the opportunity of examining biomicroscopically a case of the rarely occurring metastatic (endogenous) gonorrheal conjunctivitis associated with iritis. Zeeman (in Berens' textbook) states: "It is characterized by bilateral hyperemia of the bulbar conjunctiva and episclera; the palpebral conjunctiva is congested at times, but always in a lesser degree than the other tissues. Discharge, if present, is scanty. It may develop into subacute epibulbar conjunctivitis with multiple vesicles or papules resembling phlyctenules, and is frequently associated with vesicular keratitis or a slight infiltration of the corneal parenchyma. According to Byers (1908) 30 per cent of these cases show a diffuse opacity of the cornea, hyperemia of the iris, and opacity of the aqueous within the first few days. In fact all the signs of iritis are present."

may only partly fill the anterior chamber and tends to gravitate to its lower parts. This type of exudate forms rapidly and likewise tends to reabsorb speedily. I have seen cases in which the exudate formed and disappeared within forty-eight hours after the initial onset of the process.

In a recently observed case, about four days following its onset, in what appeared to be a simple iritis, the lower part of the pupil was suddenly occupied by a saggittally flattened-out gelatinous mass hanging suspended in the middle of the anterior chamber and, in the aqueous above it, cells and flocculent material were present. A week later the gelatinous material had absorbed and the iritis then ran an uncomplicated course for several weeks until clearing began. During this time the turbidity of the aqueous gradually became less. Keratic precipitates appeared and one permanent posterior synechia developed.

Another case occurred in a young man, aged 35 years. He had been under intermittent treatment for gonorrheal prostatitis for several years and had seven attacks of iritis during this time, four in the right eye and three in the left eye. In each eye the initial attack was typically exudative and cleared with no residua. The subsequent attacks, except the last in the right eye, were all simple and abortive, and lasted from a few days to a week with no permanent sequellae. However, the last attack in the right eye was plastic in nature and was more violent; it lasted several weeks. As a result a broad posterior iritis adhesion developed below (Plate LII, fig. 4). This synechia became permanent and even resisted such sympathomimetic agents as epinephrine and neosynephrine. The posterior corneal surface and anterior lens capsule were dusted with precipitates. There were numerous pigmented deposits on the anterior vitreous trabeculae. The vision was reduced to 20/40.

Cases have been described in which excessive cellular exudation led to hypopyon and others in which extreme hyperemia of the iris was followed by actual hemorrhage into the anterior chamber with the formation of a hyphema. These are rare occurrences. With

proper treatment recovery is the rule, although remissions are frequent, especially if the original focus is not eliminated.

The plastic or proliferative variety also may start violently and because of the tendency for the formation of broad and multiple synechiae, dire results may ensue, owing to recurrences and extension of the inflammation to the ciliary body and vitreous. The proliferations are found as whitish bands attached to, and running concentric with, the pupillary edge — especially inferiorly. A considerable amount of plastic exudate may be found adherent to the anterior lens capsule even in the pupillary area (Plate LII, fig. 3); this is seen as roundish or irregular thin, flattened, whitish plaques, some of which may be partially dusted with pigment. In the beginning numerous efflorescences may foreshadow adhesions. At times the pigment of the pupillary border is pulled off and after disintegration is scattered and deposited in the form of granules on the lens capsule, posterior corneal surface, or on the iris itself. Alternately, detached pigment may be seen in the form of ropes or strings attached to the capsule or to neighboring areas, along the pupillary margin itself. The proliferative action and fibrinous organization may result in eventual seclusion of the pupil with the attendant dangers of such a complication. Varying degrees of iris atrophy may follow depending on the severity and occurrence of remissions. although the tendency to form keratic precipitates is not a usual feature of gonorrheal iritis or iridocyclitis, successive attacks may nevertheless leave permanent fine keratic precipitates; in the extreme state their toxic effect and associated epithelial changes may cause some loss of corneal transparency. The picture of recurrent plastic gonorrheal iridocyclitis is one that in its chronic phases is clinically and biomicroscopically not specific and at times may closely resemble the exudative type of tuberculous iritis.

SYPHILITIC IRITIS AND IRIDOCYCLITIS

Although luetic involvement of the uveal tract may appear at any time following the general systemic invasion of the disease, predominantly, acute iritis or iridocyclitis occurs in the so-called secondary

stage (early or recurrent), usually in association with the skin rash and mucous patches, or as a late manifestation during the tertiary stage.* The wide variations of its incidence, as reported statistically in the literature, is probably related to differences in locale with differences in racial factors, institution of early therapy and variability in natural individual resistance.† From the standpoint of the biomicroscopic appearances all manner of variations may be found, from the early and late roseola of the iris to the marked and plastic exudative, papular, or nodular iritis. Consequently unless we meet, for example, a typical case of iritis papulosa, a specific diagnosis, by biomicroscopic means alone, may be difficult or impossible. As a general statement, luetic involvement of the iris tends to run an acute course rather than a chronic one, so characteristic of tuberculosis. Healing is usually rapid and frequently occurs with no or little sequelae, especially if treatment is instituted early.

As a matter of fact, owing to modern methods of diagnosis and especially to the rapidity and thoroughness with which treatment is applied in the early stages of infection, luetic involvement of the iris is not as common as in the past. Iritis papulosa or gumma, for instance, described frequently in the last century, are now comparatively rare.

Roseolae, which may be the first indication of systemic luetic infection, are the result of localized areas of hyperemia (Plate LII, fig. 5). Macroscopically, they appear as small irregular red spots, resembling petechial hemorrhages, but with the biomicroscope they are seen to be composed of congeries of superficial dilated vessels, probably venules or capillaries of the superficial leaf. Because of the presence of the obscuring surface pigment in brown irides, roseolae

* Acute iritis may also be initiated as the result of a Jarisch-Herxheimer reaction and, rarely, as a recidive reaction following inadequate therapy.

† For example, Igersheimer⁴⁸³ found 28 per cent of cases of acute iritis and 8 per cent of chronic iridocyclitis in syphilitics. From 6 to 70 per cent of all cases of iritis are said by various authors to be luetic. In summing up the subject of syphilitic iritis J. E. Moore states: "Whether early or late, iritis is twice as common in colored patients as in white patients with syphilis and slightly more frequent in males than in females; iritis may be expected to occur in from 4 to 5 per cent of all patients with early secondary syphilis. It is almost twice as frequent as a manifestation of a recurrent secondary syphilis, and is a fairly common manifestation of late syphilis."

are seen chiefly in blue or lightly colored irides and in these they are confined chiefly to the region of the lesser circle and ciliary zone. Deep roseolae probably occur but because of the overlying tissue are not readily visible.

Roseola iridis is an early and transient vascular manifestation, coincident with the appearance of the macular rash and mucous patches, lasting a few days and disappearing without other manifestations of iritic irritation or exudation. However, in one case that I saw, faint anterior chamber flare was present. The exact mechanism which leads to the formation of roseolae is not known. Kruckman believed that roseolae represent a mild tissue reaction to minor capillary emboli,⁵¹⁶ but pathologically, the changes in the primary or secondary stages of lues are not the result of vascular involvement as originally proposed by v. Michel (1881).⁵⁴⁴ In these stages, vessel involvement, if present, is probably secondary to changes occurring in the periadventitious connective tissue (the primary location of the inflammation) either by mechanical pressure or by extension. Experimentally, in animals Igersheimer found that vessel changes may be absent.

It is questionable whether it is proper to consider the local hyperemic areas, which occur somewhat later (up to the second year after infection) and which are associated with frank iritis, as recurrent roseolae. These lesions are more definitely part of an inflammatory and exudative reaction, and probably should more properly be called "iritis papulosa."

Savati Kowa (1927) examined a case biomicroscopically in which roseolae were found in the pupillary zone. They were situated all around the iris circumference and seemed to lie on the surface. The vessels resembled short curved rods — snakelike — with apparently blind-ending branches. Following treatment the roseolae disappeared within a few weeks. During the entire visible "life cycle" of the roseolae, there was only a minimal or faint pericorneal blush.

In a case that I had occasion to follow, in one sector the appearance of the roseolae was preceded by two radially directed vessels, running from the periphery to the lesser circle. These vessels ran

between and parallel to the iris trabeculae and, within 24 hours, at their termination in the region of the lesser circle, small roseolae formed. However, in other sectors no radial vessels were seen although there were well-formed roseolae present. Since roseolae are not accompanied by any subjective symptoms or complaints, e.g., such as pain, photophobia, tearing, or visual disturbances, it is usually only by chance that a case presents itself to the ophthalmologist.

The acute exudative and plastic forms of iritis which occur predominantly during the secondary stage (either concomitantly with the rash or, rarely, before) may also appear during the tertiary stage of the infection. Unless well-formed papules appear, those of luetic etiology may be clinically indistinguishable from the acute plastic iritis resulting from other causes. When severe, which is commonly the case, luetic keratitis is characterized by a profuse fibrinous exudation. The aqueous may be very turbid, disclosing filiform structures or even a larger gelatinous exudate in association with large, dirty gray keratic precipitates. The iris, itself, is diffusely swollen and, if lightly colored, shows hyperemic vessels arranged radially or in the form of starlike roseolae. A thin coat of fibrin may cover the iris surface. Biomicroscopically, tissue condensation within the stroma or visible elevated papules may be present.

The tendency to form synechiae is marked. These may be broad and accompanied by flat grayish exudates on the anterior lens capsule. At the height of the process the cornea usually becomes hazy not only from the presence of precipitates but also from edema of its parenchyma and epithelium. Ordinarily, the prognosis is good, especially if local and systemic treatment is instituted.

The term "iritis papulosa" is used to designate that form of acute exudative syphilitic iritis, in which elevated well-defined evanescent papules appear on the iris surface, visible by gross methods of examination (Plate LII, fig. 6). Statistics concerning the incidence of these papules in syphilitic iritis vary greatly. For instance, Groenouw⁴⁶¹ noted them in 8.85 per cent of his cases, Ozapodwasky in 23 per cent, Igersheimer⁴⁸³ in 40 per cent, and more lately Moore⁵⁴⁶ in only 6 per cent (secondary and tertiary). It should be noted that

these estimates were made for the most part without benefit of the biomicroscope. If we take into account the deeper nodular condensations within the iris stroma, which can only be properly seen with the biomicroscope (especially in the lighter colored irides), I feel that a much higher percentage would be noted.

Histologically, since the early investigation of v. Michel (1881)⁵⁴⁴, who believed that the process started within the vessels, until that of Igersheimer (1918)⁴⁸³ and others, even more recently, the tendency for the infiltrations in the uveal tract to form nodules was clearly evidenced. These nodules, which consisted of condensations of infiltrating lymphocytes and plasma cells, also contained typical giant cells but were devoid of caseation.* Deep nodular formations situated in the pupillary region may erode through the posterior iris surface and cause extensive posterior synechiae.

Iritis papulosa may usher in the manifestation of the secondary stage of syphilis, although Moore (1931)⁵⁴⁶ found it several times in a series of 109 cases of tertiary iritis. In the secondary stage it is usually accompanied by skin and mucous membrane lesions. As Zeeman has aptly stated in Berens' textbook, "As a rule, a glance at the patient's back or throat settles the diagnosis." Typically during the early stages of iritis, small round or pyramidal (Kruckmann) shaped nodules project themselves from the iris surface chiefly in the vicinity of the pupillary and ciliary margins † (Plate LIII, figs. 1-5). They vary in size from a pinpoint to 4 or 5 mm., or more. Some are solid in appearance while others look like empty cysts. Recently I had occasion to observe a case occurring in a young Negress. Coincident with a follicular-papular rash of the skin of the trunk, an acute iritis developed simultaneously in both eyes. Over the surfaces of the irides many translucent globular cysts were

* Gummas of the iris are rare and usually invade the iris by extension from the ciliary body. The essential difference between papules and gummas is evidently one of degree or intensity of involvement rather than one of fundamental structure. The resulting picture of syphilomatous infiltration (papules and gumma) is (1) formation of small papulous nodules with little tissue destruction, which heal with minimal atrophy or scarring, (2) larger nodular-like formation with extensive central ulceration (gumma and scar tissue formation), and (3) a formation, in which the process of regeneration predominates over the tendency of scar tissue.

† In about 50 per cent of cases the affection is bilateral, but both eyes may not be involved simultaneously.

PLATE LIII

FIG. 1. Papules in luetic iritis. (After Kruckmann.)

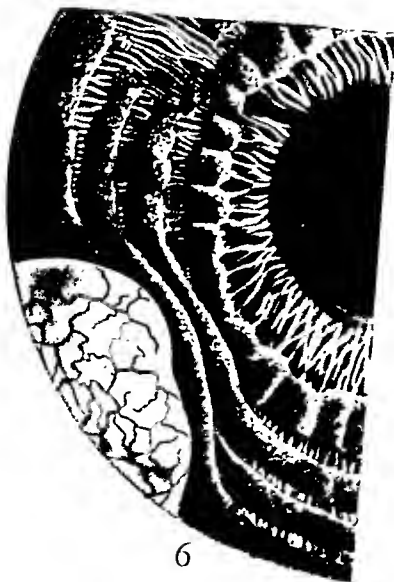
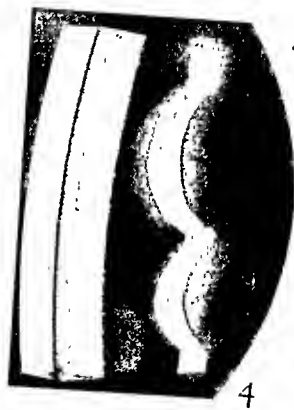
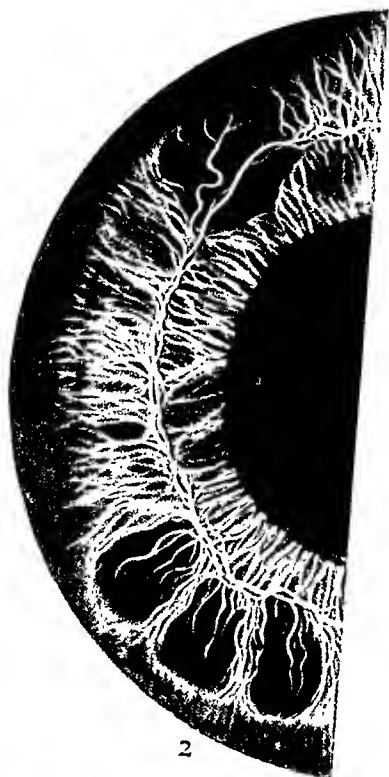
FIG. 2. Atrophic spots resulting from healed papules. (After Kruckmann.)

FIG. 3. Group of syphilitic lesions (eight months after initial lesion). Several papules and one vesicle. (After Kruckmann.)

FIG. 4. Papules, luetic iritis. Keratic precipitates.

FIG. 5. Verruca-like growth with internal hemorrhage following luetic nodular iritis. (After Kruckmann.)

FIG. 6. So-called "iris condyloma" in a woman appearing six years after initial infection.



seen forty-eight hours after the onset of the attack (Plate XLVII, fig. 5). Within a few days granules of pigment were found on these cysts. Small keratic precipitates and several posterior synechiae developed; the latter later were separated by atropinization. These floccular structures remained for about a week and then disappeared. Vigorous antiluetic therapy was begun and in about 3 weeks the lesions in both eyes quieted down; the only residua were several small pigmented keratic precipitates and pigment clumps on the anterior lens capsule.

TUBERCULOUS IRITIS AND IRIDOCYCLITIS

Since the end of the last century, tuberculous uveitis has excited the interest of ophthalmologists. With each advance in our knowledge of this condition, controversial "schools" of thought developed. Even today, in spite of the accumulated mass of clinical and experimental data, we still are forced in most instances to make the diagnosis of the presence of a tuberculous process by inference or by exclusion. In the majority of cases of endogenous uveitis, in which a tuberculous etiology is suspected, it is not always possible to recognize clinically the presence of the pathognomonic tubercle. There is no question, however, that with the aid of the biomicroscope one becomes increasingly aware of indicative iritis changes, but one must not forget that frank iritic nodules or condensations of iris tissue occur in infections of other etiologies. In many of these we can be aided by other diagnostic methods, e.g., the serology tests for syphilis, the Frei test for lymphogranuloma venereum, the agglutination test for brucellosis, etc. In certain cases of low-grade chronic tuberculous cyclitis, the iris may appear entirely normal or may show only an occasional ectodermic nodule of the pupillary margin until secondary complications (increased intra-ocular pressure or extension and progression anteriorly or posteriorly) become evident. In the early stages of this type of involvement, an aqueous flare, keratic precipitates or vitreous opacities, with or without slight ciliary injection, may be the only findings.

Excluding the forms in which clinically it is possible to make a

definite diagnosis (e.g., lues), European observers consider that tuberculosis is the chief cause of endogenous iridocyclitis, particularly the chronic type. Whether or not such a conclusion is valid, only time will prove. Already we know of forms caused by conditions which simulate the picture of tuberculosis (e.g., brucellosis and Boeck's sarcoid) and which heretofore were often considered of tuberculous etiology. Likewise there is still controversy concerning the mode of infection in tuberculous uveitis. The thesis that the uveal tract can be infected in a primary manner was developed because of the fact that tuberculous uveitis commonly occurs in apparently healthy persons with no signs of active tuberculosis; but this is no longer favored. In selected groups microscopic evidence of healed tuberculous lesions of other organs has been found on post-mortem examination in from 42 per cent (Whitehead, 1922)⁶⁷⁷ to 90 per cent (Wehlinger and Blangey 1933-1934). Since these patients died from other causes, it must be assumed that they overcame the original tuberculous infection. Considering the universality of the finding of healed or quiescent tuberculous lesions mediastinal (hilar) and mesenteric lymph nodes and localized lesions in the pleura and internal organs, it is remarkable how little tendency there is for the infection to spread. Evidently other factors must come into play. Excluding the rare acute miliary form, in which a fatal massive generalized infection occurs, it is probable that ordinarily the uveal tract becomes involved via the blood stream, following "a mild transient bacillema."

On the other hand, as has been shown experimentally in animals, the degree of ocular involvement not only depends on the "violence and massiveness of the infection" but also on the immunologic state of the animals.* Hence two main groups of reactions appear pos-

* Duke-Elder, summarizing the experimental findings of many workers,⁴⁰⁶ states: "If the animal has not been previously infected, tubercle bacilli reaching the eye produce a slowly progressive ocular tuberculosis with tubercle formation. Depending on the quantity of bacilli reaching the eye, these may be miliary or few; and depending on the virulence of the bacilli may ultimately result in caseation and perforation of the globe. If on the other hand, the animal has been previously infected, quite a different picture results from a second infection either with living or dead bacilli or even with tuberculo-protein an acute caseating lesion appears which is quite noncharacteristic of tubercle which in some cases is selflimited and heals, usually with subsequent recurrences, and in others, depending on the number and virulence of the bacilli and the resistance of the animal, progresses with an intense caseating reaction which leads to perforation."

sible: (1) the reaction to an original infection and (2) the reaction occurring in one already sensitized from a previous infection. In both instances, the severity of involvement will depend on the resistance of the patient as well as the massiveness of the infection. In the first group, if the patient's resistance is very low, acute miliary lesions may result, but if it is high, typical discrete or confluent tubercles (chronic tuberculous lesions) appear which may become progressive (destructive and diffuse) or nonprogressive (relatively benign); that is, the lesions may be typically "productive" or proliferative. The acute massive (caseating) tuberculous iritis and the acute disseminated iritis occurs in the very young and as a rule spreads to the deeper structures and results in loss of the eye. In the second group, two varieties of tuberculo-allergic reactions may appear — a chronic and recurrent exudative form or, rarely, an acute plastic form. The latter tends to affect those in early adult life while the former occurs during the fourth decade.

Although such sharp divisions, based on experimental work, may be valid for the purposes of understanding the pathogenesis of uveal tuberculosis or for establishing a working classification, clinically it is possible and probably more usual to see one form merge into another or to find simultaneous manifestations of both in the same eye. For example, in tuberculous iridocyclitis it is not uncommon to find the proliferative type of lesion (tubercle) in an iris that is also undergoing an acute plastic or a chronic exudative type of involvement. As previously mentioned, this will become more evident after such cases are carefully studied with the biomicroscope. Such confused or mixed pictures undoubtedly result from variances not only in the virulence of the organisms and their toxins but also in the patient's natural general resistance or local tissue resistance. In the same way the strict division into acute and chronic forms is only arbitrary, since even the so-called chronic variety may be initiated or punctuated by "acute" attacks or by exacerbations. Vogt, who believes that tuberculosis is the chief cause of most of the subacute or chronic endogenous anterior uveitidis, makes out a strong case for the influence that is played by heredity in the sense of an increased susceptibility. However, all persons with an inherited suscept-

ibility do not develop tuberculosis unless their resistance is lowered by exposure, poor nutrition, or poor hygienic living conditions. It would appear that this inherited susceptibility may have a predilection for certain organs (organotropism) or even certain cell groups; therefore, it is possible that an inferiority or susceptibility to tuberculosis may be transmitted by independent genetic factors not only to the whole organism but also to a specific organ or to particular parts of an organ. In order to prove that hereditary inferiority is a decisive factor in the localization of tuberculosis to even special parts of the eye, Vogt brought an impressive array of cases in which tuberculous keratitis or iridocyclitis was found in father and son, mother and daughter, aunt and niece, and in sisters. Wehlinger and Künsch have shown in a study of tuberculosis in identical and dissimilar twins and that the former (provided one of the twins is affected) is twenty times more liable to acquire tuberculosis than are the latter.

Morphologically and from the standpoint of biomicroscopy two main types appear: (1) the proliferative and (2) the exudative. In both types the process may be ushered in by an acute phase or it may start benignly with little outward inflammatory reaction and develop chronically. Also in both the ultimate outcome varies (depending on the violence of the infection and resistance of the patient) from complete resolution to degrees of damage that may result in partial or complete destruction of the eye. In the earlier literature, only the rare proliferative types were recognized, since these were characterized by the frank appearance of nodules (tuberculomas), either discrete or conglomerate, or the rapidly progressive and devastating diffuse proliferating types (also rare), in which a violent caseating and necrotizing lesion soon destroyed the entire globe. As previously mentioned, examination with the biomicroscope more and more brings to light mixed types, i.e., combined forms of the proliferative and exudative types or those which in time may merge from one type into the other.

Proliferative Types. The proliferative types are characterized by tubercle formation — a nonspecific defensive response of tissue

against the noxious agent (nodules are produced in other conditions besides tuberculosis).^{*} Depending on the resistance of the host, two processes occur within tubercles: "the one caseation — distinctive and dangerous, and the other sclerosis — conservative and healing." In tubercles of the iris both these processes occur but fortunately the severe caseating variety (e.g., diffuse proliferative tuberculosis and the destructive conglomerate tubercle involving iris, ciliary body, or choroid) are rare. These more malignant types, which tend to destroy the eye, are more common in the young. Cases have been reported in young persons in whom an acute caseating ocular tuberculosis was followed by a general body dissemination with fatal termination. In adults with higher resistance the appearance of a tuberculous nodule in the iris may occur with little or no reaction (slight or absent anterior chamber flare) and with or without any ciliary blush. Permanent and complete healing can follow, or alternately, in consequence of tissue sensitization, acute exacerbative exudative reactions may complicate the picture and lead to severe sequelae.

For the purposes of classification[†] the proliferative types of tuberculous iridocyclitis have been divided into:

- I. Acute miliary lesions
- II. Chronic tuberculomatous lesions
 - A. Benign lesions (single or multiple tubercles)
 - B. Slowly progressive conglomerate tubercles (ultimately destructive)
 - C. Rapidly progressive and destructive lesions — diffuse proliferative tuberculosis

ACUTE MILIARY LESIONS

Acute miliary tuberculosis is generally a fatal disease, in which massive and overwhelming doses of bacilli, injected into the vascular system, are widely disseminated throughout the organs. As a result

^{*} For example, syphilis, sympathetic ophthalmitis, leprosy, and as a foreign body reaction in iritis nodosa and neoplasms. Since most of these conditions can be ruled out by other methods, the frank appearance of nodules should at least be considered suspicious of tuberculosis.

[†] Modified after Duke-Elder.

PLATE LIV

FIG. 1. Tuberculous iritis. Efflorescences and one nodule at 3 o'clock.

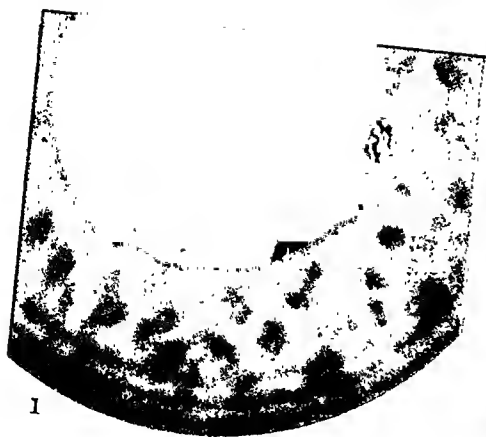
FIG. 2. Tuberculous iritis in a child.

FIG. 3. Abortive nodules in the case of tuberculous iritis.

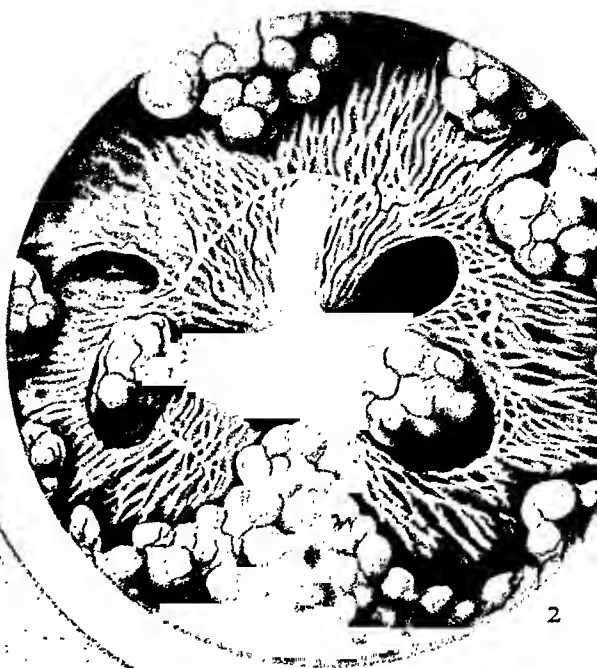
FIG. 4. Miliary tubercles and exudation in a 21-year-old man.

FIG. 5. Tuberculosis of the iris. The nodules and floccules are seen in various stages of development. Note large intrastromal nodule at lesser circle.

FIG. 6. Exudative iritis with numerous superficial floccules.



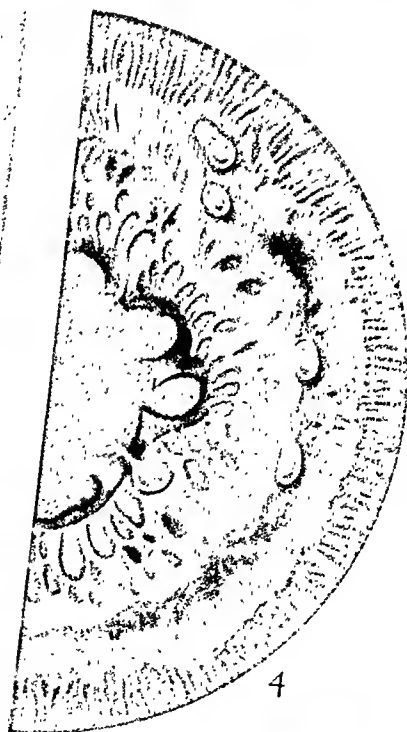
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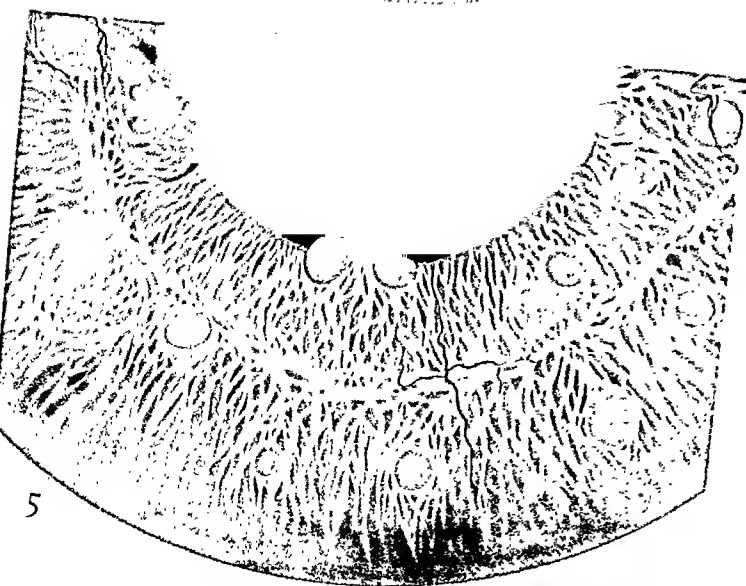
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6



4



5



the involved tissues become "shot through" with tubercles. The uveal tract, particularly the choroid, may share in the generalized metastatic involvement but clinically they have not been noticed during life as frequently as necropsy data would indicate (Plate LIV, figs. 2, 4). In acute miliary tuberculosis, the clinical presence of iris tubercles is a great rarity. The fact that they have been seen so seldom may possibly be explained by their deep location within the stroma itself. As von Michel pointed out (before the days of biomicroscopy) only superficial nodules that either elevate the anterior iris layers or project themselves from the iris surface can be grossly seen as such. Superficially-placed nodules can be seen even when they are still very small. They appear as grayish-yellow nodules, avascular and irregular, distributed over the iris surface. The more deeply situated nodules need not elevate the anterior surface of the iris but may push back the pigment epithelium layer. If not obscured by iris edema, aqueous flare, or keratic deposits the use of the narrow beam may reveal deeper condensations (tubercles) within the iris stroma.*

CHRONIC TUBERCULOMATOUS LESIONS OF THE IRIS

Benign lesions (single or multiple tubercles). In distinction to the acute miliary type, a more chronic and relatively benign form of tubercle may develop in the iris. It can occur in a quiet iritis with no ciliary injection and can be so ephemeral in character as to disappear within a week or it may be part of an insidious iridocyclitis. In more serious cases it may be preceded or accompanied by a mixed form of exudative iritis of varying severity. Rarely, as an extreme picture, confluence of the lesions may ultimately resolve into a devastating conglomerate tubercle. As already mentioned on page 830, iris nodules, efflorescences, and flocculi vary in size, location, and texture. First, there are the ephemeral efflorescences at the pupillary border, which are soft, pure white in color, and attached by a small base to the intact pupillary pigment border. (See Plate

* Frequently, the appearances of nodules may be preceded by an acute iritis of varying severity.

XLVII, fig. 4.) Second, there are the more solid mushroom-like nodules situated at the pupillary border (Plate LIV, figs. 1, 3). When these arise from behind the seam they lie on the anterior lens capsule and tend to form adhesions. With mydriasis they can frequently be separated, leaving behind pigment and exudative spots, attached to the anterior lens capsule. The surface of these nodules is granular, owing to the adherence of pigment granules and cellular exudate. Third, there are superficial efflorescences and nodules of the mesodermal iris layer (Vogt, type I). These are not a part of the iris substance but rather are attached to, and lie on, the surface. Vogt considers these as typical tuberculides. They are either more or less solidly homogeneous and opaque appearing bodies (nodules), which are best seen in light colored irides, or may resemble large glasslike efflorescences (floccules of Busacca) (Fig. 330). These structures are in themselves avascular and contain no iris tissue strands. Fourth, there are deep nodules within the stroma (Vogt, type II). These are seen as waxy exposed homogeneous nodes, pushing the iris fibers aside, as localized surface prominences covered by superficial iris stroma, or as diffuse protuberant areas of glassy thickenings (Plate LIV, figs. 4, 5). The latter, when in the neighborhood of the frill, may be easily overlooked; they are most easily detected in light colored irides. Iris vessels, if present, tend to run between or over nodules rather than into them.

Slowly progressive conglomerate tubercles. When nodules, such as described above as type II, proliferate and grow, confluence with neighboring nodules will result in voluminous masses (conglomerate tubercle), which project forward into the anterior chamber and may or may not eventually reach the posterior corneal surface. The regions of the sphincter and peripheral ciliary zone are the common places of predilection. It is possible, even after the formation of large conglomerate protruding masses, for resolution to occur with varying degrees of iris atrophy as a residual finding. On the other hand, progressive growth and extension serves to spread the disease to contiguous parts. In the periphery such formations may cause anterior synechiae by drawing the iris forward or may occlude the

anterior chamber angle. Such granulomatous masses, extending forward from the iris, may be nodulated or solid. They vary from a dirty gray or yellow to pink in color and could be compared with leucosarcoma; vessels may arborize over them but rarely invade the growths themselves. This picture becomes readily evident on employing the optic section. Nodular masses at the root of the iris may result from extension of a progressive conglomerate tubercle of the ciliary body. Invasion of the sclera via the drainage system (spaces of Fontana and Schlemm's canal) is evidenced by a scleroperikeratitis (sclerosing keratitis). The surface of the iris may also be inoculated from conglomerate tubercles of the ciliary body by the implantation of tuberculous material. Loosened particles from the inner surface of the ciliary body carried by the fluid currents become attached to the iris. Corneal affection may result from direct contact with masses in the anterior chamber or after perforation of the sclera. Tuberculous granulations, spreading over the cornea, cause a tuberculous keratic ulcer, which may result in perforation. Although the ciliary body tends to block posterior extension, deeper involvement via the pupil has been reported. With the formation of exudate, the pupil may be secluded and occluded. Extension to the lens may result in spontaneous rupture of the lens capsule and its involvement (described histologically by Mayou in 1903⁵³⁸).

The rapidly progressive and destructive form of diffuse proliferative tuberculosis is fortunately a rare occurrence. It is characterized by a highly toxic and necrotizing reaction which destroys all the ocular tissues. The eye becomes acutely inflamed and painful. The iris is thickened and heavy masses of exudate fill the anterior chamber. Posterior extension produces a picture similar to that seen in purulent panophthalmitis.

PLASTIC AND EXUDATIVE TUBERCULOUS IRIDOCYCLITIS

A tuberculous etiology for a nondescript plastic or exudative iritis or iridocyclitis of tuberculous origin can in most cases be inferred only by exclusion of all other etiologic factors. In a limited number of such eyes, tuberculosis has been demonstrated histologically. Mild

and self-limited cases of iritis and iridocyclitis do not come to autopsy, and even in the more severe ones, in which complications necessitate removal of the eye, one should never make a positive clinical diagnosis without reservation. Among the features in the clinical aspects of a case which should be considered in this connection are (1) an insidious onset, (2) prolonged duration with a tendency to exacerbations, (3) the amount and character of the exudate (especially the keratic precipitates and the exudate on the anterior lens capsule), (4) the appearance (especially late) of localized condensations within the stroma, and (5) the degree of destruction in the iris itself.

The fact that tubercle formation occurs in the iris after the onset of a plastic or exudative iridocyclitis is well known to those who have carefully studied and followed these cases biomicroscopically. Whether nodules or nodular swellings on or in the iris indicate true tubercles is still unknown. As Vogt suggests (he examined such lesions histologically in one case), they may represent the tuberculid in the sense of Darier.

Both the *acute plastic* or the subacute or chronic types of iridocyclitis may start as a simple endogenous inflammation i.e., with an anterior chamber flare and slight ciliary injection. In the plastic type, within a few days this progresses into a highly inflammatory process, characterized by marked ciliary injection, pain, and considerable blurring of vision. Efflorescences at the pigmented pupillary border or seam presage the formation of posterior synechiae. The turbid aqueous frequently reveals the presence of larger flocculated material as well as dotlike cells. Keratic precipitates and varying degrees of edema of the corneal epithelium and stroma will be noted. The sudden appearance of a deep infiltration in the region of the limbus (sclerosing keratitis), which sometimes occurs, is in my opinion a diagnostic sign of great importance and, after ruling out other granulomatous infections, probably pathognomonic of tuberculosis.

The visibility of iris vessels (particularly the radiating ones) is most pronounced in light colored irides. Typical nodules (charac-

teristic of the proliferative forms) are not seen as a rule. Because of the nonspecific character of plastic iridocyclitis, it usually is impossible to make a definite etiologic diagnosis by the clinical picture

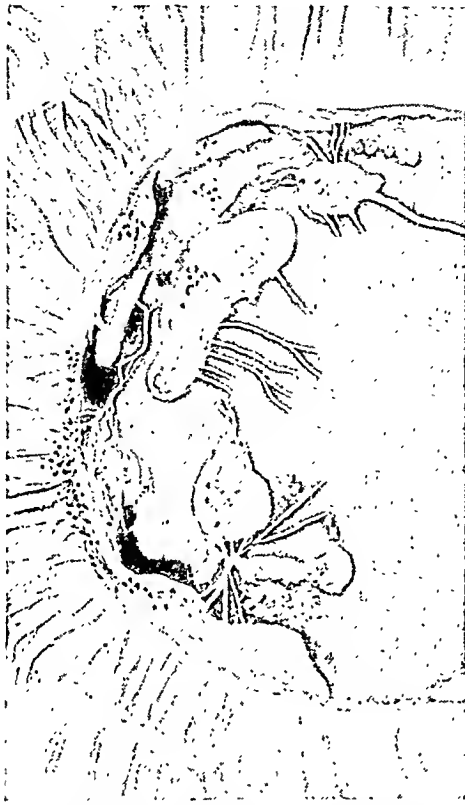


FIG. 333. Chronic type of tuberculous exudative and productive (combined form) iridocyclitis. (After Vogt.) Note exudative membranes at pupillary margin and tubercles on anterior lens capsule.

alone, especially in the initial attack. Recurrences may, however, result in pictures that resemble the chronic exudative or relapsing form, ordinarily seen in older individuals.

The chronic types of exudative iridocyclitis, which start more insidiously and are characteristically relapsing, are clinically more suggestive of a tuberculous origin. They may be ushered in with little, if any, ciliary injection but are soon followed by a progressive and relentless exudative process. As a result, the posterior surface of the cornea becomes studded with large precipitates of the "mutton-fat" type. Broad and multiple synechiae may eventually lead to occlusion of the pupil. Flat exudative membranes may extend over the anterior lens capsule (Fig. 333). These at times actually pull

stromal tags over the obscured or missing seam. These tags (dentate in type) become firmly attached to an exudative membrane on the lens capsule. The iris, which rapidly becomes atrophic, changes in color and results in the well known "sick-eye" appearance. It is in these cases that the biomicroscope will reveal the atypical nodular swellings mentioned above. Cellular infiltration in the vitreous usually occurs early but this together with the lens changes soon obscure any view of the fundus. Such a process may quiet down at any stage or may ultimately lead to loss of the eye.

Tuberculous Cyclitis. As indicated before, every iritis is probably associated with reaction in the ciliary body. Especially in the more severe forms, the exudative and proliferative changes within the ciliary body contribute in a large measure not only to the severity of the process but also to the ultimate fate of the eye. In the fulminating forms involvement of the iris masks the process occurring in the ciliary body; although direct inspection is impossible, involvement of the latter may safely be assumed. This of course has been borne out by histologic study and even clinically by evidences of complications, e.g., changes in intra-ocular pressure and vitreous opacities. Conversely, lesions which start as a primary focus in the ciliary body (page 808), may, by forward extension, involve the iris. Perhaps the most common form of cyclitis is the one that begins quietly with few signs of inflammation and that subjectively causes only a slight blurring of vision. Biomicroscopic examination at this time reveals only a faint flare in the aqueous and the so-called "mutton-fat" keratic precipitates. Such a reaction may remain minimal and with reabsorption leave no trace. I had occasion to see such a case in the left eye of a colleague, aged 37, who had one recurrence in 3 years. Each time a slight blurring of vision and a degree of tenderness to palpation were noticed. There was no definite ciliary injection at any time, and the highest intra-ocular tension recorded during both phases was 28 mm. Hg (Schiötz). The attacks of blurring, which lasted about a week in each instance, were associated with definite flare in the aqueous and with several discrete large "mutton-fat" keratic precipitates. A few white cells were seen in

the anterior parts of the vitreous. There was no visible hyperemia of the iris or efflorescences at the pupillary margin. Examination one month after the last flareup revealed complete resorption of the precipitates. The fundus was entirely normal. An extensive medical survey gave negative findings, except for the radiographic diagnosis of an old healed pulmonary lesion.* This case illustrates a form of "quiet" tuberculous cyclitis which, without the aid of the biomicroscope, might easily have been overlooked. In others, the symptoms may be more marked and progressive, e.g., efflorescences of the pupillary margin with or without synechiae, hyperemia of the iris, a more pronounced infiltration in the vitreous, and increased intra-ocular tension. Secondary glaucoma is a formidable complication, which in the end may destroy the eye.

LEPROSY OF THE IRIS

Barros † in his excellent monograph has classified the iritis of leprosy clinically as follows: (1) Diffuse acute, subacute or chronic form, serous or serofibrinous; (2) localized form: miliary or nodular leprosy.‡

Acute Diffuse Iritis. Acute diffuse iritis of leprosy presents a fulminating serous or serofibrinous type of iritis characterized subjectively by pain, blurring of vision, photophobia, lacrimation, and blepharospasm. Only rarely are both eyes involved simultaneously. Occasionally it develops by extension from an episcleral lesion,§ i.e., close to the limbus. Objectively the findings differ little from the acute violent iritis resulting from other causes. Ciliary injection is marked; corneal edema, bedewing, and keratic precipitates are present. In the aqueous the changes, depending on the amount and character of the exudate, vary from a simple Tyndall flare in the beginning to the formation of a large coagulum, not unlike that seen in gonorrheal iritis. In addition to the larger coagula, numerous threads may be seen,|| which at times may assume a cobweb form.

* This patient's mother had died of tuberculosis at the age of 29.

† Barros, from the standpoint of biomicroscopy, has written the first authoritative description of ocular leprosy, "Some Clinical Aspects of the Ocular Involvement in Lepers."

‡ The term "nodule" is used in the morphologic sense rather than in the true histologic one.

§ See Volume I, pages 219 and 515.

|| See Volume I, Plate XL, fig. 3.

At the height of the inflammatory process there may be an increase in intra-ocular tension. Synechiae are common and often intractable (Fig. 334). Barros only observed the presence of a hyphema on two

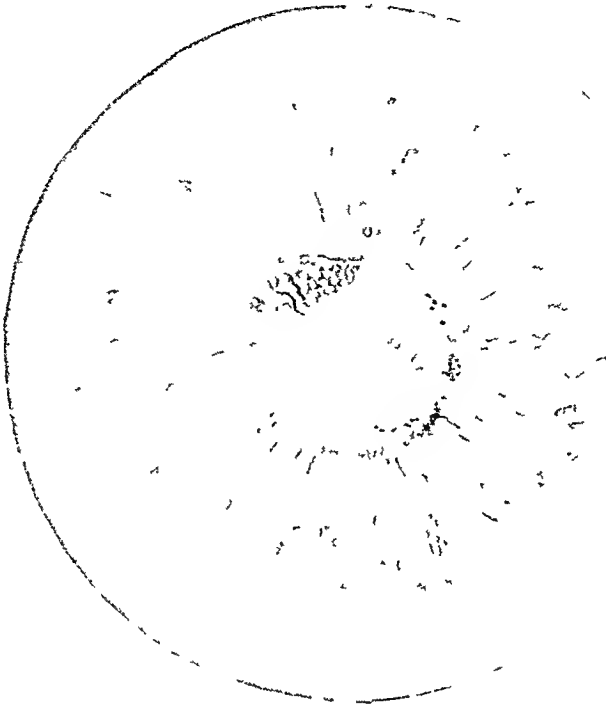


FIG. 334. Acute diffuse iritis of leprosy. (After Barros.)

occasions but hypopyon-like masses are frequently observed in the bottom of the anterior chamber. These at times may be somewhat obscured by an abnormally relucant limbal spur. As is the case with all types of acute fulminating iritides all manner of complications and sequelae may result: iris atrophy with generalized or localized areas of depigmentation, seclusio pupillae, secondary glaucoma and in the extreme cases, phthisis bulbi.

Subacute or Chronic Diffuse Iritis. The reaction in some of these cases may be so mild as to be distinguishable only by the biomicroscope. Subjectively the visible congestion may be absent or minimal and the principle symptom, if elicited, may be varying degrees of blurred vision, secondary to corneal edema and aqueous flare. In these cases both eyes may be involved simultaneously. The flare (exudation in the anterior chamber) may be present for long periods and there is less tendency to the formation of synechiae as compared with the acute type. In some the keratic deposits may be fine, but

at times in others the larger mutton-fat type of deposit may be found. The exudate accumulated in the angle may become organized. The accumulation of any considerable amount of exudate on the posterior corneal surface exerts a toxic effect and frequently is followed by a secondary keratitis. Exudations of this kind at the pupillary margin may, after dilation and rupture of a synechia, leave a mass of pigment or a grayish nodular mass on the anterior capsule of the lens. Barros also described two cases in which trauma initiated a subacute attack of iritis in lepers. In both these cases the iris was covered with pearllike nodules (combined form). The fact that trauma may bring on an attack of leprous iritis is reminiscent of similar episodes in cases of hereditary lues in which ocular trauma may cause an acute attack of interstitial kerato-iritis.

Miliary and Nodular Iritis of Leprosy. This type is signaled by the appearance of numerous pearllike structures, commonly situated in the deeper iris layer between the frill and the pupillary border, in other words in the sphincter region. These efflorescences or pearls are best seen in light colored irides by indirect illumination. Often there may be a complete absence of iritic irritation or inflammation (no flare). The granulations may be present for years with no acute symptoms. Hence, Barros states that "the eyes have a good tolerance to them." Generally the condition is found to exist bilaterally with no interference with vision, although the pupillary reaction may be sluggish and dilation difficult. If not fully developed, the presence of these nodules may be overlooked unless indirect illumination with an overloaded lamp, and high power of magnification ($60\times$ or more) are employed. When numerous, Barros has described their appearance as like a necklace surrounding the pupil (Fig. 335). They also may fill the crypts and later on may be found in the whole iris. In some cases with nodules of the deep mesodermal layer, transpupillary retro-illumination of the iris may reveal holes of the posterior retinal iris layer. Barros stressed the point that these nodules must be differentiated from the more whitish and superficial "pearls" of exudate seen in the diffuse serofibrinous forms. These may also be found in the pupillary area and may develop into large

irregular cauliflower masses. The latter are superficially placed on the iris surface while the miliary "pearls" are located within the iris thickness. In the early stages they can only be seen by indirect

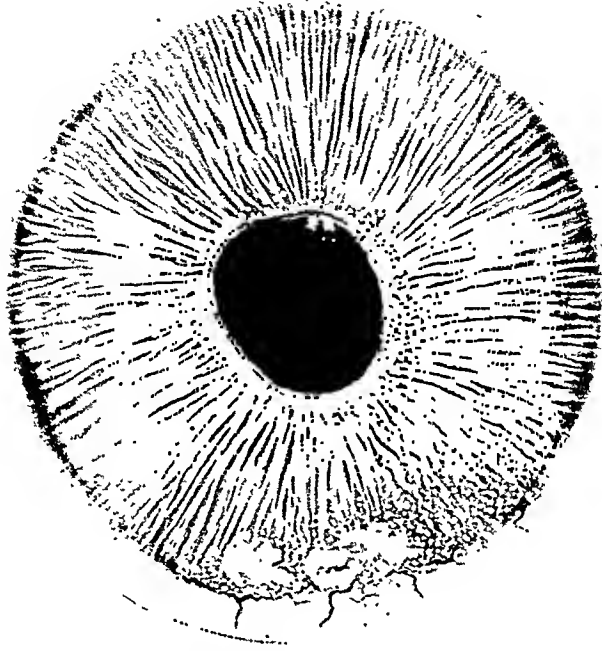


FIG. 335. Miliary and nodular iritis of leprosy. (After Barros.)

illumination but later are visible by direct focal light after they become larger and more superficially placed. Barros states that he was unable, even after years, ever to see the transformation of these miliary lesions into the more conspicuous lepromatous nodule, although this should not be impossible since the latter is considered to be a miliary leproma.

As regards nodular leprosy of the iris, Barros in his large series only observed it in one case; in which in the right eye a large mass in the iris tissue extended from the iris root at 9 o'clock gradually decreasing in size to the pupillary margin. There was considerable exudation in the aqueous and a large keratic precipitate extending near the limbus from 9 to 6 o'clock.

It should be pointed out that both exudative diffuse iritis and miliary nodules may at times be found simultaneously or consecutively in the same eye.

HETEROCHROMIC IRIDOCYCLITIS

In this section only that form of complicated heterochromia, which is commonly designated the "heterochromic syndrome of Fuchs," will be considered. The so-called simple hereditary type of heterochromia has previously been mentioned in the chapter on iris coloration (page 768). Likewise color changes resulting from atrophic states of the iris following senility and inflammations, glaucoma (vitiligo), etc., which some writers consider heterochromia, have also been discussed. The rarer so-called "neurogenic form," secondary to a sympathetic lesion, presents nothing of special biomicroscopic interest (although Herrenschwand⁴⁷⁰ claimed that in this type he noted no atrophic changes in the deeper iris trabeculae).

The syndrome of heterochromic iridocyclitis is characterized by the presence of a unilateral asymptomatic low-grade process which is devoid of congestion or pain and is slowly progressive over a long period of years. Among the important features are the change in iris coloration (as contrasted with that of the unaffected eye), the mutton-fat keratic precipitates, and the complicated cataract. The color of the iris in the diseased eye is always lighter than that of the unaffected eye. The disturbance in coloration, which usually precedes the other findings, is diffuse and at times may be very slight. Because of incorrect impressions of color that may be gained when the iris is inspected with a highly focalized light, as is obtained with the biomicroscope (page 764), it is preferable, especially when the color change is slight, to make comparisons between the two eyes in daylight. This is especially true when both irides are blue. The affected iris loses its fluffy character, the trabeculae (in light colored irides) appear stretched, flattened and as if they were bound together by a gelatinous material. Occasionally a radial trabecular vessel may be seen. Especially noteworthy, in addition to the atrophy of the stromal melanophores, is the depigmentation of the posterior retinal pigment layer of the iris. Transpupillary retro-illumination will practically always reveal depigmentation of the pupillary margin,

not unlike that of ordinary senile atrophy, disclosing radial streaklike or cuneiform patches of depigmented areas. Similarly the remainder of the posterior pigmented iris surface may be affected so that retro-illumination will disclose multiple streaks or innumerable pinpoint holelike areas giving the iris a typical motheaten appearance. At times, owing to marked atrophy of the stroma, the posterior pigment layers will "shine" through, and the iris of the diseased eye may appear darker than the normal fellow eye (especially if it is blue or gray). Posterior synechiae are a rare occurrence in heterochromic iridocyclitis. However, Vogt found a small glassy type of nodule at the pupillary border (and only rarely elsewhere) on or in the iris. The presence of such nodules in this condition has not been mentioned in histologic studies of the iris, nor did Vogt have the opportunity of studying them histologically. The finding of iris nodules would seem to strengthen the theory of infection as the etiologic factor in heterochromic cyclitis, especially the tubercle, as opposed to the hypothesis of Fuchs who postulated the action of toxins in early or intra-uterine life. Often the pupil of the lighter colored affected eye, due to changes in the sphincter, is slightly dilated and reacts sluggishly to light.

The keratic deposits are of the mutton-fat type, sometimes round, sometimes star-shaped, and usually situated in the lower parts of the cornea. In either case their edges are sharply delineated, the "smeary" or plaquelike forms not being found. In spite of the marked iris depigmentation the keratic precipitates remain white or gray; a fact that distinguishes them from the precipitates of other conditions which tend to become dusted with pigment, especially as they age.

Complicated cataract is common (Plate L, fig. 5). The opacification usually begins in the posterior cortex in the form of punctate dots. However, in contrast to the usual "cataracta complicata" (page 1167) the typical granular opacity with iridescence at the region of the posterior pole does not appear. Both nuclear cataract and anterior cortical changes soon follow so that eventually the whole lens becomes opaque. However, in the case of a man, 50 years

of age, the type of cataract as it developed could not be distinguished from the ordinary senile variety. Operation for cataract extraction in these eyes is well borne.

Biomicroscopic examination of the anterior parts of the vitreous, when it can be seen, frequently reveals the presence of white dots adherent to the fibrillar structure of the framework. Pigment dust in the vitreous in these cases is a great rarity. Likewise it is very unusual to find the denser and larger forms of opacities in the vitreous.

Glaucoma, a serious complication, usually appears late in the disease and may prove to be very stubborn.

SYMPATHETIC OPHTHALMITIS

The type of reaction that occurs in sympathetic ophthalmitis, both in the exciting eye and in its sympathizing eye, is similar clinically in many respects to tuberculous exudative iridocyclitis. In practically all cases the exciting eye shows or gives a history of a perforating injury in the ciliary region or of a previous operational site. Mixed forms of sympathetic ophthalmitis are seen in which the exudative process is accompanied by a proliferative one in the form of nodules or efflorescences. Generally the injured eye fails to heal properly; it remains irritable and the inflammatory process, either acutely or gradually or with recurrent flare-ups, develops into a typical chronic exudative iridocyclitis. There is a tendency for the aqueous flare to remain permanent even during periods when the lesion is relatively quiescent. Keratic precipitates and varying amounts of corneal edema soon form (Fig. 336). The iris becomes thickened and discolored, and pupillary efflorescences are followed by broad posterior synechiae. Localized iris thickenings in the form of deep nodules may form with progressive exudative membranes extending from the iris to cover the lens capsule. Dispersed pigment granules incorporated in the exudate are frequently found. Radiating iris vessels tend to grow over the pupillary edge and lie in the membrane which is adherent to the capsule. As a result of interference with its metabolism, the lens becomes cataractous. Seclusion

of the pupil may lead to increased intra-ocular pressure but as a rule concomitant involvement of the ciliary body, choroid, and vitreous and the formation of cyclitic membranes (with retraction

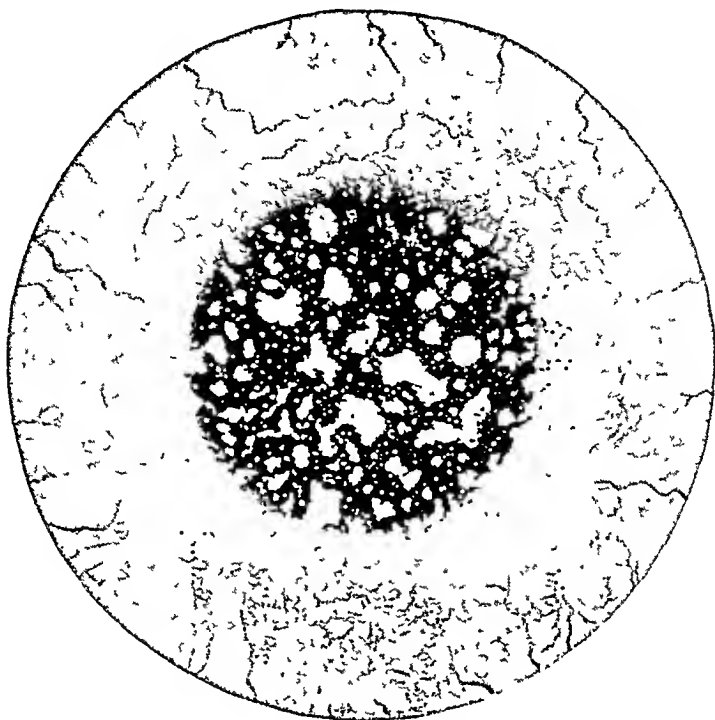


FIG. 336. Sympathetic ophthalmia. (After Gilbert.)

of the wound) results in hypotension, obliteration of the anterior chamber, atrophy, and eventually phthisis bulbi.*

From the standpoint of biomicroscopy our interest in the sympathizing eye lies chiefly in the early signs that warn us of its involvement.†

* As Duke-Elder has pointed out: "While this clinical picture of a low grade, recurrent traumatic uveitis with tendency to phthisis is typical, it is by no means invariable—for sympathetic inflammation may follow almost any type of condition—on the one hand a clean wound that has apparently healed rapidly and without complications and on the other a violently acute irido-cyclitis, or even on rare occasion a purulent panophthalmitis."

† There are many references in the early literature concerning sympathetic irritation, apparently a reflex condition. In the presence of a badly diseased or injured eye the fellow (healthy) eye may suddenly become irritable as evidenced subjectively by slight conjunctival injection, photophobia and even neuralgic pains. Objectively no positive signs of real intra-ocular involvement are found. Several years ago I had occasion to encounter such a reaction recurring in an eye, in the case of a young man 34 years of age, in which the opposite eye was rendered useless by several fulminating attacks of tuberculous uveitis over a period of six months. Glaucoma developed consequent to intractable seclusio pupillae and posterior sclerotomy and cyclodiathermy were resorted to. Eventually this eye became painless and atrophic. Several months later the patient began to complain of "eyestrain," tearing, slight blurring and occasional photophobia in the healthy eye. Careful examination revealed only a questionable injection of some of the bulbar vessels. The tension and visual fields

When confronted with an injured eye, especially when a perforating wound involving the ciliary region has occurred, *careful and frequent biomicroscopic examinations of the fellow (uninjured) eye are mandatory*. It is only by means of the biomicroscope that we are able to detect a faint aqueous or vitreous flare (see Vol. I) with or without the presence of cells. In the first few days the signs may be so slight that they may not evoke enough blurring of vision to be noticed by the patient subjectively. But one could not make a clinical diagnosis of sympathetic ophthalmitis from a complaint of visual disturbance alone, since even from the very beginning the presence of exudation in the chambers of the eye is the *sine qua non* of the diagnosis. However, as the exudation increases the patient usually becomes aware of it.*

As the flare in the aqueous becomes more turbid, cells and larger flocculi can easily be observed in the outstanding beam. By this time keratic deposits will be found. Small at first, they rapidly increase in number and size until eventually typical mutton-fat precipitates

were normal. The tarsal conjunctiva was normal. Refraction demonstrated that the eye was emmetropic. Biomicroscopic examination showed no evidence of corneal edema, aqueous flare or cells in the vitreous. A mild collyria was recommended. With the use of the collyria and an application of silver nitrate 0.5 per cent once a week, the symptoms abated. A month later there was a recurrence of the "irritation" and after two more recurrences I advised that the diseased eye be enucleated. Pathologic examination of this eye revealed the residua of a nondescript exudative iridocyclitis, secondary glaucoma and general atrophy of the globe. Since this was done (four years ago) the patient has had no further irritations in his good eye. Evidently it is possible to have a "sympathetic irritation" which is not necessarily followed by genuine sympathetic ophthalmitis.

* Children, for example, may have considerable blurring of vision without complaining about it; this holds true also for unintelligent adults. One would like to be able definitely to answer the question as to whether at the very outset, cellular infiltration or deposits on the framework of the vitreous occur before they appear in the aqueous cells. Only once, in a case six weeks following a cataract extraction, was I able first to see white cells deposited on the structure of the vitreous in the fellow (unoperated) eye. The patient (a diabetic) had an extracapsular operation on the left eye which was followed by low grade iritis. At the time I saw the patient in the clinic the operated eye was still inflamed and he complained of slight blurring of vision in the opposite eye. Under mydriasis the eye which had no ciliary injection showed a few white cells just behind the lens and deeply on the fibers of the vitreous. I was unable to detect an aqueous flare, even after becoming well dark-adapted and using strong illumination. Ophthalmoscopic examination was negative. However, two days later, the flare in the anterior chamber was marked and ciliary injection was present. Within two weeks a typical plastic iritis developed. Following fever therapy and high doses of salicylates the reaction in both eyes subsided, but the lens in the sympathizing eye rapidly became cataractous. Some months later an attempted de Wecker discission on the left eye resulted in a violent reaction which eventually led to phthisis bulbi. This eye was enucleated. A year later the right (sympathizing) eye was operated on for cataract. This likewise was followed by a devastating reaction which ultimately ended in a shrunken globe.

develop. Owing to the convection currents, the effect of gravity and the condition of the endothelium, they tend to be disposed in the well-known inverted triangular shaped figure in the lower cornea. Edema of the corneal endothelium occurs early. According to Vogt, transparent droplets can be brought into focus at this level by retro-illumination. As is usual in all types of iridocyclitis epithelial edema appears somewhat later, particularly as the general inflammatory reaction increases. Cases have been reported in which the reaction abated at this stage, i.e., cells in the aqueous and vitreous and keratic deposits disappeared and recovery followed. In other words the entire reaction in the sympathizing eye may at times run a short course, similar to a mild cyclitis. In the same way, in spite of relapses of a mild nature extending over a period of years, serviceable vision may be retained. Unfortunately such happy outcomes are not the rule. Following the initial signs of exudation in the chambers of the eye, the condition either rapidly or chronically passes into the state of a progressive devastating iridocyclitis, which in most respects is similar to the picture seen in the severe exudative types of tuberculosis. The iris becomes hyperemic, discolored and thickened. Efflorescences form at the pupillary margin and together with exudative membranes which extend from the pupillary margin over the lens capsule lead to broad synechiae and ultimately to seclusio pupillae. In addition to efflorescences, nodules (described by Vogt) in or within the stroma may occur similar to those in tuberculosis. Iris vessels are commonly present and, with the formation of exudative capsular membranes, these extend over the pupillary area of the lens. Opacification of the cornea following epithelial and endothelial edema (best seen by retro-illumination), together with precipitates and secondary stromal opacities, may hinder a clear view of the deeper structures. In the same way cataractous changes may preclude inspection of the posterior segment. Increased intra-ocular pressure attends blockage of the aqueous circulation in the anterior chamber either after seclusio pupillae (with or without iris bombé) or after peripheral anterior synechiae.

In fulminating cases, exudation may be so great as to form a

progressive hypopyon of such proportions that absorption becomes impossible. Organization may result in the appearance of a large, solid, dirty yellowish mass filling the entire anterior chamber. Such an eye stays chronically inflamed for months and either remains hypotonic or, after passing through a phase of hypertension, is hopelessly destroyed.

ESSENTIAL PROGRESSIVE ATROPHY OF THE IRIS

According to Ruby, about 35 cases of essential progressive atrophy of the iris have been reported in the American literature. Clinically, this condition is characterized by the appearance of a unilateral * progressive atrophy of the iris, in which marked shrinkage and retraction of the tissue tends to form large colobomatous defects. Eventually most of these eyes are destroyed by absolute glaucoma. The condition starts with an eccentric position of the pupil, due perhaps to the fact that "the pupil would naturally be drawn to the side where the iris is less atrophic." McKeown⁵³⁰ noted this in the case which he reported; the pupil was first vertically elongated upward and was later displaced downward, being dragged by the less atrophic iris before the progressive atrophy advanced into the upper temporal quadrant. Most of the reported cases occurred in early adult life and only in rare instances was the diagnosis made before the onset of glaucoma. The pupillary margins may show varying degrees of ectropion of the retinal pigment. High degrees of atrophy appear in the unretracted portions of the iris, leading to the formation of "holes," which are well demonstrated by diaphanoscopy and by retro-illumination. Troncoso, who made a gonioscopic examination in McKeown's case (Fig. 337), 14 years following the onset of the process, found several anterior peripheral synechiae. This was the first case of essential progressive atrophy of the iris in which gonioscopy was employed in the examination of the angle of the anterior chamber. However, it is hoped that in the future,

* Fine and Barkan⁴²⁷ reported a case occurring bilaterally in a boy of nine, who at the age of 5 years was first brought for examination because of a deformity of pupils. Several operative procedures for control of tension failed; in one eye detachment of the retina occurred and in the other, a definite buphthalmos.

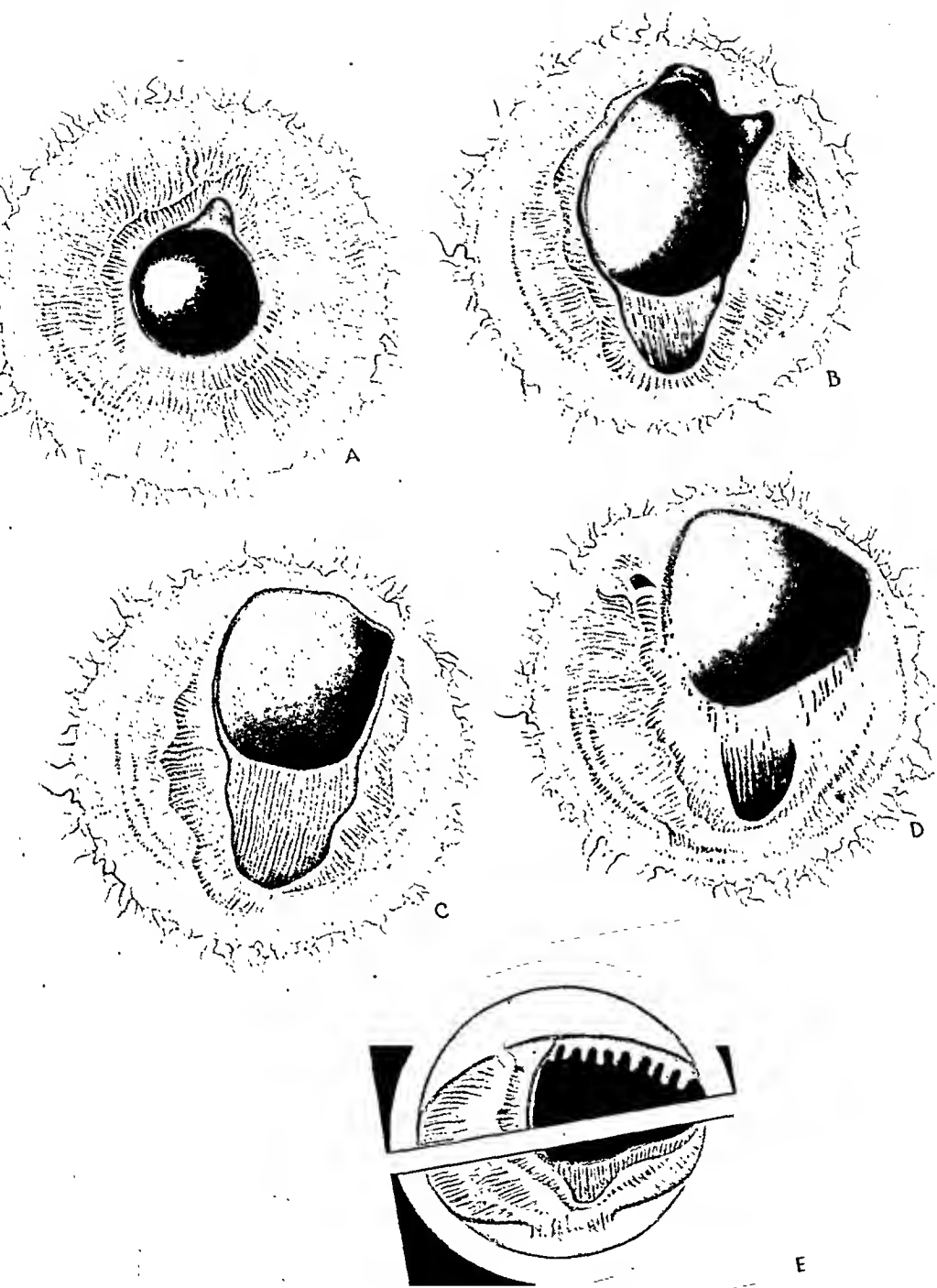


FIG. 337. Essential progressive atrophy of the iris. A-D, progressive stages of the disease; E, view of angle by means of gonioscope (late stage). (After McKeown.)

studies of the angle will be made at the very beginning of the condition in order to determine whether the disease starts there or whether the formation of anterior peripheral adhesions is a later secondary manifestation. The etiology of essential progressive atrophy is obscure. The absence of frank signs of inflammation would indicate a degenerative process, and its late appearance possibly abiotrophy. Von Grosz suggested that it is a primary degeneration on a hereditary basis.⁵⁸³

POLYCYTHEMIA VERA (VAQUEZ-OSLER DISEASE)

In contrast to engorgement of the retinal, choroidal, and conjunctival veins seen in this disease, congestion of the iris vessels occurs rarely. This may result in a reddish discoloration of the iris, especially noticeable in light colored irides. Chamber hemorrhage in this condition has not been reported. Varying degrees of iris atrophy with pigment dissemination may follow. Tirrelli has reported on the fact that he noticed a fine dustlike pigmentation unevenly distributed between the lesser circle and the root of the iris.

LYMPHOMA

Rarely as part of a generalized acute or chronic leukemia, infiltration of the iris with white cells may occur either in the form of massive infiltration or as circumscribed single or multiple tumors. Several authors have described isolated cases of leukemia in which the iris became affected but in which the pattern of involvement differed. Three forms were noted: (1) the nodular type in which yellowish tumors appeared on the iris surface. These resemble tubercles and during the active stage were more or less vascularized; (2) the diffuse type in which the iris assumes a grayish thickened appearance and in which the markings and trabecular relief become obliterated. This form may be complicated by the appearance of hypopyon (Weve); (3) the hyperemic form, characterized by extensive engorgement of the iris vessels followed by bleeding into the anterior chamber with the formation of hyphemia. Bab³⁵³ described a case of leukemia in which a typical exudative iritis was noted.

Chapter Twenty-Two

TRAUMATIC LESIONS, CYSTS, AND TUMORS OF THE IRIS

TRAUMATIC (MECHANICAL) LESIONS

FOLLOWING an injury to the eye, the iris alone may be affected, i.e., it may be the only part of the eye that reveals any residual damage; but, of course, other ocular structures may also be affected.

Needless to say, the type or extent of a traumatic iris lesion depends on the force and direction of the blow, the character of the insulting instrument, and whether or not the injury was a perforating one. In the absence of gross alterations, the biomicroscope is of incalculable value in revealing, especially in retrospect, the presence of minute iris changes attributable to trauma. This may be of great importance in medicolegal work and particularly in industrial or war injuries. We shall not dwell on the gross injuries or those in which (owing to severe corneal or scleral lacerations or perforations) large portions of the iris as well as other intra-ocular tissues (ciliary body, lens, vitreous, etc.) may be damaged or even extruded. This discussion will be confined to those types of traumatic iris lesions, which, owing to their delicacy, may be overlooked by the ordinary methods of examination.

Injuries, such as contusions in which no perforation has occurred but in which the force of the blow transmitted to the iris is sufficient to cause damage to it, are sometimes known as "indirect" traumatic lesions. These may result in iritis, dislocations, and ruptures of iris trabeculae, tears in the sphincter with notching of the pupillary margin, traumatic mydriasis, dehiscences of the retinal pigment layer and iridodialysis. As an immediate consequence of any injury to the iris we may find hemorrhage in the anterior chamber and/or

within the iris itself (interstitial), occasionally general dispersion and precipitation of pigment on the boundary walls of the anterior chamber and transient exudative pupillary membranes (fibrinous). (See Vol. 1, page 572.) The conception of an anterior segment traumatic syndrome was advanced by Frenkel ⁴³¹ and was confirmed by Davidson, ³⁹² who studied a series of 34 cases of contusions and analyzed their sequelae. The syndrome has to do with relatively minor sequelae, the recognition of which depends to a large extent on the application of biomicroscopy. Not all the sequelae are found in every case, but the regular appearance of two or more of these changes, which are anatomically related, give credence to such a conception. The changes seen in the iris consisted of traumatic iridoplegia, small iris tears, dehiscences of the posterior retinal pigment layer of iris, radial or concentric, single or multiple, with especial predilection for the iris periphery (root). The combination of iris dehiscences as seen by transillumination under mydriasis, in which sectorially a straight line chord replaces the normal arclike curve of the pupillary margin is, as Davidson states, incontrovertible evidence of a contusion involving the anterior segment. The chord-like appearance of the pupillary edge which is similar to that seen in frank iridodialysis suggests a minimal rupture or paresis of the dilator in which the sphincter is also involved. Other sequelae of the anterior segment traumatic syndrome are found extending backward from the iris root along a pathway which predominantly involves the lens equator, zonule, anterior vitreous, ciliary body, and ora serrata. These will be referred to again in the special chapters on some of these subjects.

Direct traumatic lesions to the iris are those in which the iris tissue is wounded by direct contact with an instrument or foreign body and hence can only occur with a perforation of the cornea or sclera. The distinction between direct and indirect injuries depends on whether or not evidence of a perforation can be found. Actually as far as the iris itself is concerned nearly all the alterations found in indirect injuries also may occur after direct ones.

Traumatic Iritis. Severe direct trauma to the eyeball, either with perforation (e.g., by a sharp instrument, a foreign body or by operative means), or without perforation, may be followed by varying degrees of acute iritis or iridocyclitis. This is evidenced by the presence of an aqueous flare owing to the outpouring of albuminous exudate and cellular elements. The cellular elements derive from the blood absorb relatively rapidly. Fibrinous coagula in the forms of pupillary membranes may appear and disappear within several hours.* Rupture of the iris tissue itself is usually accompanied by hyphema. Undoubtedly the presence of chamber exudates, fibrin, blood, and pigment excites iritic irritation. That the presence of a highly albuminous fluid or product is toxic and may induce iritis in some sensitized persons is not surprising. It is well known that this occurs frequently in retinal separation or in the presence of degenerated lens material in the anterior chamber following extracapsular cataract extraction or ruptured traumatic cataract. As in nontraumatic iritis, hyperemia of the iris, seen best in light colored irides, is manifested by increased visibility of the blood-carrying iris trabeculae, and, at times, of circular coursing vessels. Synechiae may form, and depending on the severity of the process, varying degrees of iris atrophy may follow eventually.

Iris atrophy following traumatic iritis (in the absence of tears) may in nowise differ from that seen after ordinary iritis and may be manifested by general or localized areas of stromal vitiligo or by areas of increased translucency seen by retro-illumination or by transpupillary diaphanoscopy. Alajma noted the appearance of radial lacunae, especially in the ciliary zone, by retro-illumination (transpupillary diaphanoscopy). In severe cases in which trauma is followed by prolonged iridocyclitis associated with either low or high intra-ocular pressure, the eye may eventually be lost.

On abatement of the acute phase biomicroscopic examination generally shows the presence of pigment precipitates on the posterior corneal surface, the iris, the lens surfaces, and in the vitreous. The pigment (hematogenous and uveal) tends to disappear rapidly from the anterior chamber or from the chamber boundaries (especially

* See Volume I, page 573.

from the posterior corneal surface). This is particularly true with contusions uncomplicated by chronic iritis. Examinations of such cases months or years after the accident generally fails to demonstrate any trace of pigment on the posterior corneal surface. On the other hand pigment dispersed in the vitreous tends to be lasting.

Safar described a case in which pigment deposited in the contraction folds of the iris resulted in the appearance of pigmented curved lines on the surface of the iris. I have noticed at times in traumatic iritis that the adhesions (synechiae) are composed of whitish tags — evidently exudate. Particularly in the young, migration and secondary diffuse deposition of pigment granules on the iris surface may darken it so that by comparison with the uninjured eye the iris of the injured eye becomes darker, resulting in a sort of heterochromia in reverse. The genesis of the traumatic pigmentation of the superficial iris layer has not been solved. Clinically it is indistinguishable from the ordinary pigmentation of the normal brown iris. Discoloration of the iris from blood pigment is yellowish in appearance and follows hemorrhage in the anterior chamber or in the iris stroma itself. Vogt suggests that the stimulus of the trauma or even inflammation can cause a new formation of pigment by the superficial layers of the iris.

This type of posterior traumatic heterochromia may not appear until considerable time has elapsed. Koby cited a case in which this occurred about 10 years after the injury while the interval of development in Vogt's cases was from 20 to 30 years. Posttraumatic heterochromia has also been recorded after operations for congenital cataract, especially needlings.

Ruptures and Tears of the Iris. Following direct contusions to the eyeballs, even without perforation, ruptures and tears of the iris tissue are very common. These may be so slight as to be overlooked unless the iris is carefully studied with the biomicroscope.

Tears and ruptures of iris tissue are usually followed by hemorrhage. Owing to the effect of gravity the blood sinks to the bottom of the anterior chamber, giving rise to a hyphemia. The lower or older part of the hyphemia becomes darker than its horizontal

PLATE LV

FIG. 1. Partial rupture of the iris at the pupillary border. Traumatic cataract.

FIG. 2. Rupture of the iris at the pupillary border exposing the deeper posterior leaf.

FIG. 3. Incomplete hole in the iris following penetrating intra-ocular foreign body.

FIG. 4. Linear rupture through the iris substance.

FIG. 5. Traumatic separation of the anterior leaf of the iris.

FIG. 6. Traumatic cyst of the iris.



border. When the focal beam is passed through the anterior chamber above the solid part of the hyphemia, a typical Tyndall flare will be observed, caused by the suspension of yellowish or reddish brown cells. Owing to the density of the cellular material little or no convection currents will be noted.*

As a rule the absorption of blood following chamber hemorrhage is rapid. However, cellular deposits on the posterior corneal surface and visible aqueous flare may be present for some time, but even these tend to absorb rapidly. As soon as the blood begins to settle and to absorb, it becomes possible to inspect the iris and to evaluate the damage incurred. Where slight displacements of the pupil and irregularity in its contour are present, especially when associated with a twisting or separation of the trabecular iris system in one segment or another, trauma should be suspected.

When small, ruptures of the sphincter appear as minute radial notches in the pupil. Larger ruptures, which may even extend to the ciliary region, may be visible with the unaided eye and are differentiated from coloboma by the fact that no pigment excrescences are seen along the pillars of the defect (Plate LV, figs. 1, 2). However, ruptures of the iris do not necessarily have to include the sphincter nor do they always extend through all the iris layers. They may only involve the stroma in varying locations, degrees and depths or the posterior iris face alone. An example of partial rupture of the stroma is seen in those cases in which only the frill itself is torn. Minimal ruptures or separations of the posterior iris layers involving the dilator muscle in which the sphincter may also be affected (without any visible notching of the seam) results in the characteristic D-shaped pupil. In these, the defects in the posterior iris layers may only be visible by strong transscleral or transpupillary diaphanoscopy. These dehiscences are found in the sector corresponding to the chord-shaped defect in pupillary contour and generally at the iris periphery. This type of defect of the pupillary contour is seen best in iridodialysis. Traumatic dehiscences are more liable to occur at the periphery where the iris is thinnest. By diaphanoscopy or

* See Volume I, page 577.

retro-illumination they appear reddish in color. They may be single or multiple. The smallest ones may be seen as faintly glowing dots. Others, larger, are round, irregular or radiating, not unlike the atrophic lacunae seen in senility or iris atrophy (glaucoma, postinflammatory, etc.).

Tears of the sphincter *per se* also result in changes of pupillary contour. Even the smallest tears which result in a minimal notching will cause moderate dilation of the pupil. This will be permanent. Unless bridged by inflammatory exudate torn edges of iris tissue do not cicatrize. Consequently separations in iris tissue, whether traumatic or operative, are permanent. Multiple radial lacerations of the sphincter and larger tears correspondingly result in greater pupillary dilation and deformation.

Faulty action of the sphincter may cause folds by releasing the tension of the iris fibers, and in the same way some torsion of the adjoining fibers results. This torsion of the fibers adjacent to the torn area is characteristic; they assume a corkscrew aspect, in contrast to the normal radiating and somewhat stretched appearance seen with the pupil undilated. Partial or complete separation of the superficial stromal leaf from the deeper stromal layer has been reported many times after injuries. When extensive this may simulate the so-called "senile detachment." The detached superficial portions may float freely in the anterior chamber and noticeably do not follow movement of the iris. As a consequence the detached parts do not move with dilation of the pupil.

Holes in the iris, secondary to perforating injuries, particularly with the passage of small foreign bodies into the deeper parts of the eye, may be missed unless transpupillary retro-illumination is performed (Plate LV, fig. 3). They should always be looked for especially when a scar traversing the corneal layers is found. Occasionally some depigmentation of the iris surface (atrophy) will be noted in the immediate neighborhood of a hole. Since most perforating foreign bodies strike and enter the eyeball obliquely iris holes occur more commonly in the ciliary zone.

Iridodialysis or separation of the iris from the ciliary body like-

wise may follow nonperforating injuries as well as perforating ones. It practically always is attended with marked chamber hemorrhage and frequently is not noticed until the blood is absorbed. As in the case with severe iris ruptures, pigment dispersal results.

Depending on the injury, an iridodialysis may be so slight as to appear as a linear black crescentic slit at the periphery of the iris, seen with difficulty and at times only when the patient's eye is moved peripherally in the direction of the lesion so as to permit the observer to inspect the far angle of the chamber. Such extreme peripheral tears are best studied gonioscopically. In iridodialysis the pupillary margin in the affected sector always slopes off to form a straight line. Larger separations permit the view of the lens equator and the zonule. With these the iris is thrown into lax folds running more or less parallel to the separated iris edge.

Interstitial Iris Hemorrhages. Ruptures and tears of the trabecular tissue of the iris is usually accompanied by chamber hemorrhage. However, bleeding within the iris substance itself may occur. When this happens, there may be no outward indications of its presence when the iris is viewed by diffuse or direct focal illumination. The blood confined to the deeper iris layers can only be seen by indirect illumination. Observation of the iris adjacent to the part illuminated by the focal beam, will reveal a reddish glow. Unless looked for carefully soon after the injury this type of suggilation or interstitial hemorrhage of the iris can easily be overlooked. In a case which came under my observation in which suggilation occurred in a lightly colored iris, examination 6 months after the trauma revealed a slight darkening of the iris as compared with the uninjured fellow eye. The superficial stroma of the iris assumed a yellowish brown hue, evidently a form of permanent blood staining. Von Hippel stated that a long standing hemorrhage in the anterior chamber and in the vitreous as well may result in a form of hematogenous siderosis. In most instances, however, blood is rapidly absorbed and leaves no trace. According to Wolff⁶⁸⁰ the importance of the iris in the absorption of a hyphemia is illustrated in chamber hemorrhages following an iridectomy. The blood disappears rapidly over

the iris but may remain a much longer time in the area of the coloboma. He showed an illustration of a histologic section of iris from an eye which was removed 24 hours after a perforating injury, in which the blood on the front of the iris is entering it by one of the crypts. The phagocytic function of the iris is probably of considerable aid to the main drainage system (Schlemm's canal) in cleansing the anterior chamber not only of blood but also of other noxa.

Interstitial iris hemorrhages may involve a considerable portion of the iris stroma or may be localized as small ecchymotic spots. When searching for such spots one moves the focal beam slowly across the iris face, looking the while at the unilluminated portions on either side of the brightly illuminated part. A hemorrhage will glow slightly reddish in the somewhat darkened areas in the faint light of indirect illumination. As soon as the beam itself falls over the area of the hemorrhage, this reddish glow is lost since it is obscured by the brilliant illumination and reflexes of direct focal illumination.

SIDEROSIS OF THE IRIS

Following the introduction and retention of a ferrous foreign body within the eyeball, within a certain time (latent period — from a few weeks to years) definite changes may occur. Owing to chemical changes the iron is converted into a diffusible form and is disseminated by the tissue fluids (especially the aqueous and vitreous). It then stains, according to Wolff, all the structures with which it comes into contact, so that all structures bathed by aqueous and vitreous will be affected. As noted with vital staining (see Vol. I), highly differentiated cells (epithelium, nerves, etc.) take up dyes readily. Also the phagocytic action of epithelial cells is well known. It is not surprising then in siderosis that the iris (anterior border layer), ciliary epithelium, lens epithelium, and retina (especially ganglion cells and neuro-epithelium) are first stained. After being taken up by these cells, the iron is probably reconverted into an insoluble form. Eventually through interference with metabolism the affected cells degenerate.

The toxic effect of siderosis within the globe is illustrated by the accompanying iridocyclitis. This varies in severity but usually is indicated by the presence of a long standing aqueous flare. Concomitantly bedewing of the corneal epithelium, endothelium and keratic precipitates occur. When this is found in conjunction with a yellowish to brownish discoloration of the iris surface (as compared to the fellow eye), siderosis should be suspected. If added to these there is a perforating corneal or scleral scar and the characteristic rust stain below the lens capsule (page 1281) (the lens soon becomes cataractous) the diagnosis is almost certain. Histologically in the iris, the staining (Perl's reaction) is most marked in the anterior border layer and in the sphincter and dilator muscles. Involvement of the latter may account for the commonly occurring incomplete mydriasis. The diffuse rusty discoloration of the siderotic iris differentiates it from the normal pigmentation of an unaffected brown iris (Plate LXXVI, fig. 2). The color change in siderosis of the iris depends on the original iris coloration and on the degree of involvement. In the case of a blue or gray iris it may change to greenish yellow or even light brown. If the iris originally was brownish then it may become darker. At any rate its color will certainly differ from that of the unaffected fellow eye. Especially important is the fact that the normal trabecular structure of the iris becomes indistinct, the iris surface becomes flattened, and there is a peculiar loss of luster — a manifestation of iris atrophy. Posterior synechiae to a cataractous lens exhibiting reddish-brown rust stains, completes the picture. Transpupillary retro-illumination of the iris at this stage is not always possible but transscleral illumination will reveal the typical lacunae of iris atrophy.

CYSTS

The biomicroscope affords unusual means of viewing and studying cysts of the iris. When confronted with a swelling or bulge in the iris, our first concern is to determine whether the structure is solid or cystic. In the case of a cyst, indirect illumination (i.e., placing the beam to one side of the lesion) will in most cases reveal its semi-

transparent nature. A similar effect can be obtained by viewing the iris indirectly by a modified form of scatter. The beam is directed to the corneoscleral junction, and the swelling in the iris is then observed while oscillating the beam to and fro over the cornea for a distance of from 2 to 3 mm. Sufficient light is reflected to transilluminate the involved area of the elevated iris (Plate LV, fig. 6). The importance of demonstrating this translucency will easily be understood when examining dark iritic masses, especially when a differentiation between a pigmented cyst of the posterior retinal epithelial layers and a malignant melanoma is necessary. Given an iris cyst, it is not always possible to make an etiologic diagnosis biomicroscopically, but it may be possible by exclusion to narrow down the probabilities. The biomicroscope will aid, for example, in deciding whether or not inflammatory signs, i.e., keratic precipitates or synechiae, etc., or evidences of trauma (corneal scars, ruptures of the iris, dispersal of pigment, alterations in the lens) or of a previous operation (coloboma or aphakia, etc.) are present. In an adult, only in the absence of a history of trauma or surgery or of any of the above signs might one be justified in the early stages in diagnosing a cyst as of spontaneous origin. In most instances the wall of the cyst is thin regardless of etiology or location. As a cyst develops, the iris trabeculae of the superficial layers that form anterior limiting layers of the cyst are pushed aside and become atrophic. This leaves a white, yellowish, or pearl-gray smooth surface with a characteristic silky gloss. Continued growth of the cyst tends to separate the atrophic mesodermal layers from the posterior pigmented layers. In such cases the posterior pigmented layer may even become the posterior wall of the cyst and after bulging backward it may even extend into the posterior chamber. There, in certain instances, it can be seen as a dark globular mass in the pupil. These types of dissection are destructive. The iris adjoining the cyst may exhibit varying degrees of atrophy, but more commonly it is pushed aside and folded. These folds are concentric to the rim of the cyst. Such stretching and pulling causes displacements and distortions of the pupil. At times, larger cysts may actually cover the pupillary

opening, partially or completely. The lack of resistance anteriorly permits easy expansion in this direction, so that as the cyst develops, it eventually comes in contact with the posterior corneal surface.

Meeting the resistance of the cornea it adjusts itself to the curvature of the posterior corneal surface and may ultimately cause corneal opacification. Posterior extension may result in further iris atrophy, luxation of the lens, and cataract. If the cyst is not destroyed (by removal, by aspiration and injection of a sclerosing solution, or by diathermy, etc.), loss of the eye from glaucoma or secondary inflammation may in time occur.

As Duke-Elder has pointed out, the classification of cysts on an etiologic basis is uncertain, since in most instances (excepting only the traumatic and parasitic cysts) it is not possible to explain their occurrence. With this in mind he has classified them from the clinical standpoint as follows: *

- I. Congenital cysts
- II. Parasitic cysts
- III. Traumatic implantation cysts
- IV. Exudative and degenerative (secondary) cysts
- V. Spontaneous or idiopathic cysts
 - A. Stromal (epithelial, endothelial)
 - B. Cysts arising from the pigmentary epithelium

CONGENITAL CYSTS

Theoretically two types of congenital iris cysts are possible, but clinically and even histologically it may be impossible to differentiate between them: (1) stromal cysts, apparently caused by implantation and growth of surface epithelium, corneal or conjunctival, within the stroma of the iris and (2) those derived from the posterior pigmented (retinal) epithelium. The former, similar to dermoid cysts of the conjunctiva (described by Straeten and v. Duyse⁶²¹) may start as small blebs within the iris stroma and may grow to a large size, eventually forming large semi-opaque globes

* Gilbert⁴⁵⁰ has classified them as (1) implantation cysts, (2) cysts of the iris stroma, and (3) cysts of the pigment layer; while N. Teulieres and I. Beauvieux⁶³⁵ have designated them (1) traumatic cysts, (2) parasitic cysts, and (3) spontaneous cysts.

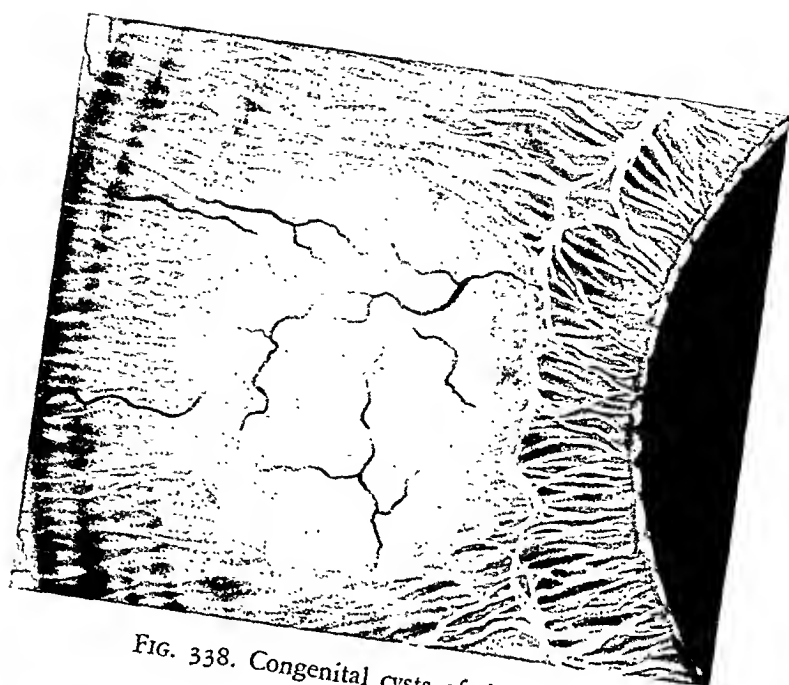


FIG. 338. Congenital cysts of the iris.

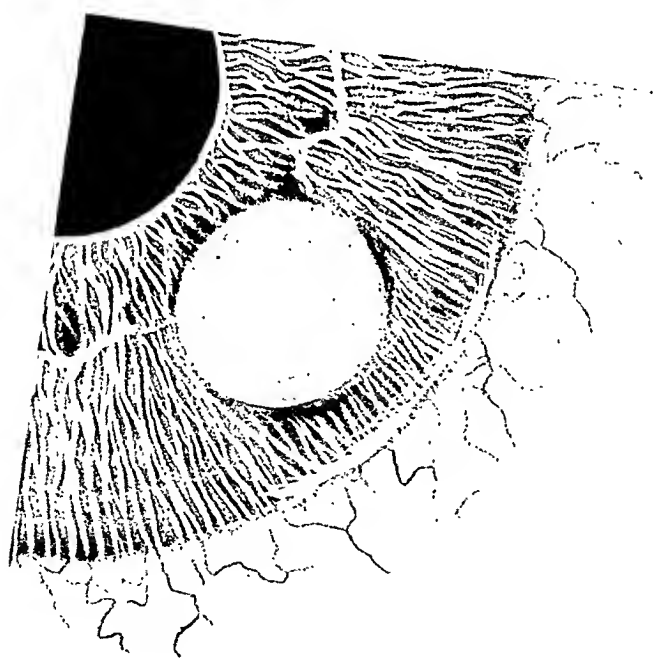


FIG. 339. Epidermoid cyst (pearl cyst).

extending into the anterior chamber. (See Fig. 338; Plate LVI, figs. 1, 2.) The second variety may result from failure in fusion of the two primitive layers of the optic vesicle (those of the pupillary margin [e.g., see flocculi, page 787] from lack of closure of the marginal sinus) or they may be derived from cells detached from the posterior pigment layer or from those in direct continuity with it. Normally, derivatives of these cells form the musculature of the iris (page 733) and also the clump cells of Koganei. Aberration in development of these migratory cells may result in the formation of cysts.

PARASITIC CYSTS OF THE IRIS

Cysticercus. Invasion of the iris by the cysticercus loa has been reported several times. The cyst appears as a transparent bubble varying in size, the interior of which may contain a whitish opaque nodule — the actual head of the parasite. Characteristic movements of the parasite within the cyst distinguishes it from other forms of cysts. At times the parasite may emerge from the cyst. If it is not removed, a violent plastic iridocyclitis may develop. This is frequently complicated by hypopyon and eventually complete opacification of the cornea.

TRAUMATIC IMPLANTATION CYSTS

Two forms of traumatic implantation cysts have been described: ⁶⁷⁹ (1) the so-called "solid" or "pearl" tumors or cysts (epidermoid) and (2) true serous cysts (translucent). These types of iris cysts form as a consequence of implantation epidermal cells (especially in association with implanted eyelashes) or of surface epithelium, corneal or conjunctival, either as free particles or as a continuous ingrowth between the lips of perforating surgical or traumatic wounds. It should be remembered that the term "pearl cyst" is correct only morphologically, since histologically, degeneration of its contents (fatty or cholesterol changes) may result in the formation of a translucent cyst, indistinguishable even with the

PLATE LVI

- FIG. 1. Congenital cystic-like mass in the iris periphery.
FIG. 2. Congenital cystic-like mass with deformity of the pupillary curvature.
FIG. 3. Benign mass of the iris (congenital).
FIG. 4. Nonmalignant melanoma of the iris. (Diffuse illumination.)
FIG. 5. Same case as Figure 4, viewed by direct focal illumination.
FIG. 6. Tumor of the iris. Nonmalignant. (After Messmann.)



1



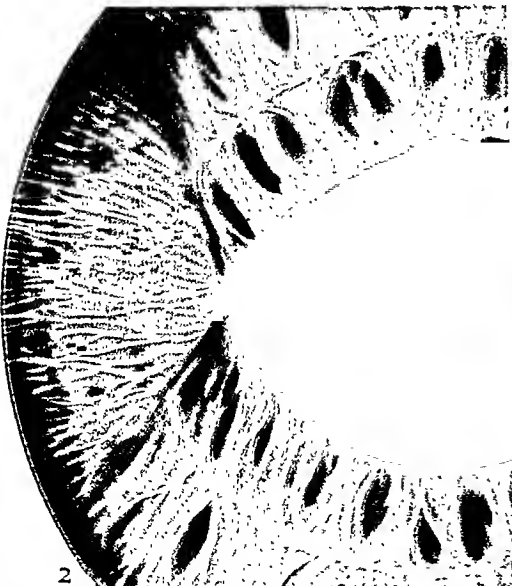
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6



4



2



3



biomicroscope from the true translucent ones.* Hence strictly speaking, the designation "pearl cyst" clinically refers to cysts whose surfaces have a dull, gray luster, comparable in appearance to that of a "black" pearl (Fig. 339). They may be either epidermoidal (which begin as solid growths) or of a true serous character (from the implantation of non-keratinized corneal or conjunctival epithelium).

It is usually only in the early stages following a perforating injury when the presence of implanted lashes in the eye can be seen, that it is possible from the morphologic standpoint to diagnose a cyst as truly epidermoidal in character.

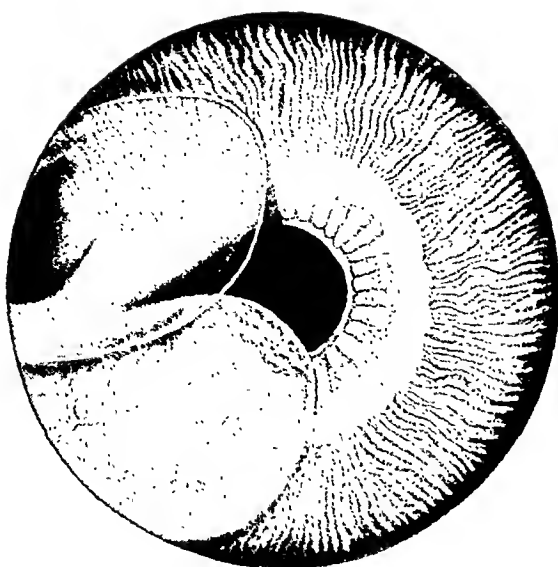
Epidermoidal Tumors or Cysts ("Pearl"?). Epidermoidal tumors or cysts start as more or less solid, whitish nodules within the iris or on its surface (Fig. 339). Their color and smoothly reflecting mother-of-pearl surface differentiate them from structures of different origin (true neoplasms). The biomicroscope frequently demonstrates that their walls are made up of concentric layers. Unless liquefaction occurs the contents are solid. Histologically these contents consist of a waxy substance admixed with cells more or less degenerated. These cells are chiefly epithelial in origin since apparently it is only epithelium that will survive and grow when implanted into the anterior chamber. The tumor consists of a cover of epithelium enclosing the waxy core mixed with cells; in the center there is a lash which is the cause of the new growth.

A large percentage of them have been seen in children and young people following perforating injuries by sharp instruments (scissors, knives, etc.). Buhl and Rothmund (1872)³⁷⁵ first explained their origin. In most instances pearl cysts of the iris followed the accidental implantation of eyelashes to which small pieces of epidermis were

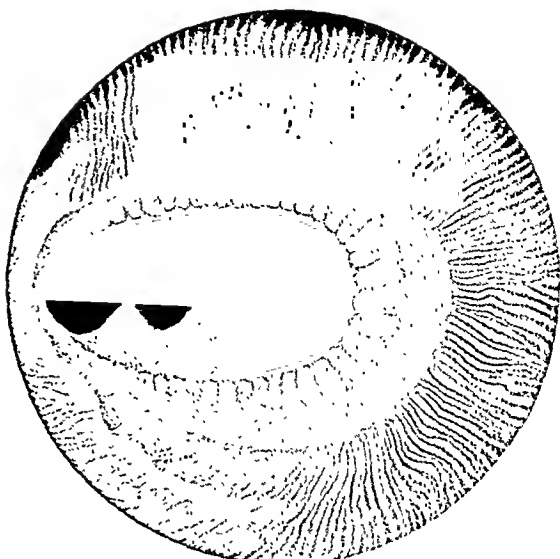
* Pathologically real pearl cysts consist of keratinized lamellae. Such "hornlike" changes would indicate that they originate from implantation of skin rather than from corneal or conjunctival epithelium. These epidermoidal cells probably arise from the root of the cilia. Silva (1905)⁶⁰⁸ has emphasized that only the cilia of the young have germinal cells attached to their roots, and that the physiologic growth of these cells occurs when they are implanted in the iris. The shaft of the lash itself may be lifted away from the iris by the growing cells which attach its root to the iris. According to Gilbert, the iris plays only a small part in the genesis of a true pearly cyst. The iris tissue forms a fertile media for its growth and helps to form its walls.

attached. In recent cases in which the cyst is already developing the eyelash may be visible clinically but with time bleaching of the pigment within the eyelash causes it to resemble a white line (Wintersteiner). It usually takes from three to six months for the cyst to develop. Its growth is not steady but is characterized by periods of inactivity alternating with periods of rapid growth and secondary irritation. These cysts may attain great size, occupying a sector of the iris and extending from the chamber angle to the pupillary margin, projecting forward until they reach the cornea. Blondel described a cyst the size of the lens filling the anterior chamber; this cyst required six months to develop. Eventually, such cysts interfere with drainage and cause increase in intra-ocular pressure; a complication subjectively attended with pain, inflammation and visual failure.

True and Serous Cysts. By far the greater number of iris cysts fall into this category. These are the ones commonly seen following perforating injuries and operative procedures (Plate LV, fig. 6). Those following injuries are generally found connected or related to a corneal scar near the limbus (Fig. 340 C). They are usually thin-walled, may be uni- or multi-ocular and often have an irregular sausage-like shape. In diffuse light, depending on the amount of residual iris tissue covering them, they may have a characteristic appearance so much like a "black pearl" that in my opinion at times they deserve the name "pearl cyst" even more than do the epidermoidal cysts (described above). In some cases the dull pearly sheen may be masked by the presence of whitish lines over the surface, the remains of stretched iris trabeculae. In such cases, it will often be seen that at the peripheral edges of the cyst, these fibers are pigmented (in brown irides), and gradually merge into the surrounding iris stroma from which they are derived. By pressure, the adjacent iris stroma is frequently thrown into concentric folds. These cysts, which so often start at the periphery, tend to grow in a concentric manner; but owing to lesser resistance in the anterior chamber they soon bulge forward and commonly cover the pupillary opening in part or completely (Fig. 340 A, B). Cases have been seen histologi-



A



B



C

FIG. 340. A. Double cyst of the iris. B. Same cyst after operation. C. Cyst of the iris. Note corneal scar. (After Buonacolto.)

cally in which the cyst was attached to the anterior lens capsule. Parsons⁵⁵⁸ described a case in which the cyst formed in the posterior chamber. The anterior wall of the cyst was formed by atrophic iris, which was completely adherent to the posterior surface of the cornea, the posterior wall of this cyst was bound by an epithelial layer that covered and was attached to the ciliary processes and the lens. On reaching the posterior corneal surface, to which it may become adherent, the surface of the true or serous cyst flattens out and lateral extension may ensue. Examination with the narrow beam generally reveals the walls of the cyst and the transparent slightly opaque fluid content. The anterior surface, indicated in optic section as a line, will have a brighter yellow color while its posterior boundary (retinal pigmented layer) is dull brown. As previously mentioned, posterior dissection and extension into the posterior chamber may occur with or without extension into the anterior chamber. In the latter case the cyst may be seen through the pupil as a dark, smooth globular mass.

In epithelial cysts of the anterior chamber, especially those following cataract operations (when the epithelium grows down between the lips of the wound), the epithelium-covered iris surface forms the posterior wall of the cyst. Biomicroscopic inspection at the site of the wound may reveal vessels passing posteriorly through the entire corneal or limbal thickness. The ingrowing tracts of epithelium extend into the chamber angle over the iris. Since epithelium is less reflective and relucient than the corneal stroma (see Vol. I), such tracts will appear dark as compared to the grayish opalescence of the surrounding tissue. Histologically the posterior wall of the cyst lies on the iris surface and is composed of parchment-like epithelium. Endothelial (traumatic) cysts of the iris are rare and follow direct trauma to the iris itself. The lumen of the cyst is lined by the endothelium of the iris surface or what appears to be mesodermal iris tissue. It usually follows folding or pleating of the iris associated with extensive anterior and posterior synechiae and consequently the liquid contents of such cysts is derived from the aqueous. In the same manner, retrocorneal cysts, in which proliferation of the cor-

neal endothelium is a factor, may enlarge and contact or become incorporated in iris tissue. Extension of the ingrowing epithelium along a corneal wound in connection with anterior chamber cysts may form large pericorneal cysts which in the end may become larger than the entire globe.³⁶⁶

EXUDATIVE CYSTS OF THE EPITHELIAL LAYERS

Separation of the retinal layers of the posterior surface of the iris by fluid or exudate is the result of stasis after inflammation; it may also result from conditions in which there is a severe interference with the iris circulation, as in glaucoma, or as a consequence of sudden lowering of the intra-ocular pressure (e.g., from paracentesis). Fixation of the deeper layers of the iris epithelium to the lens or to a cyclitic membrane undoubtedly also aids the development of such a separation. The secretory and proliferative property of these cells and the resultant tendency to cyst formation has already been noted in the discussion of the cystic flocculi (page 786). The anterior bulging of the iris stroma of such a cyst formation may resemble iris bombé. I recently saw a case in which a large posterior iris cyst developed after discission of a secondary membrane. The stroma of iris, which was only slightly compressed, protruded forward on the temporal side almost touching the posterior corneal surface. It was impossible to transilluminate the cyst's contents or to demonstrate a cavity by the optic section. However, with a hypodermic syringe it was possible to withdraw about $\frac{1}{2}$ cc. of a clear straw-colored fluid. Without withdrawing the needle itself a similar amount of 2 per cent tincture of iodine solution was injected. This caused a slight ballooning of the cyst. In a few weeks it could be seen that the walls had adhered to one another. Wilmer⁶⁷⁸ observed a case of chronic uveitis in which numerous small pedunculated cysts were seen at the pupillary border. Frequently these cysts of the epithelial layers of the iris are found histologically in eyes badly crippled from chronic inflammatory disease and glaucoma. In a case reported by Collins there was a malignant melanoma of the ciliary body.

SPONTANEOUS CYSTS OF THE IRIS

In rare cases, cysts of the iris are found whose etiology cannot be explained either by the history or by the objective findings. These cysts may either be stromal in type or may develop from the posterior epithelial layers. The fact that these cysts are seen most commonly in the young (without signs or a history of trauma) would strongly suggest the presence of a developmental anomaly. It is difficult to explain how or when a migration or implantation occurs of the several different types of cells which line the stromal type of cyst. It is easier to explain the formation of the spontaneous cysts of the posterior epithelial layers since embryologically these layers are separate. Failure in fusion from some cause during fetal life seems to be a logical explanation for the formation of such cysts.

TUMORS

The benign nevi and melanomas are discussed on pages 766-767. Here we shall briefly consider leiomyoma, angiomas, the malignant melanoma and carcinoma.

LEIOMYOMA

There have only been about eight cases of this rare type of iris tumor reported in the literature (Fig. 341). Clinically it is indistinguishable from malignant melanoma and it has been diagnosed only histologically with special stains that disclose myoglial fibers. In only three instances has the diagnosis been verified pathologically (Verhoeff, 1923; Frost, 1936⁴³² and Ellett, 1939⁴¹⁶). This type of epiblastic tumor probably arises from the sphincter muscle. Frost⁸⁰ in describing his case, states, "it was a small circumscribed nodule, yellowish-gray in color, the surface near the pupillary border being dotted with pigment. There was also an ectropion of uveal pigment at this point, producing a somewhat pear-shaped pupil. This ectropion was 1 mm. in width and showed the radial columns of dark brown pigment of the posterior surface of the iris. It suggested that

the iris had been turned inside out by contracture of the tissues in the anterior layers. The growth appeared to have increased the thickness of the iris about 1 mm. and extended from the pupillary

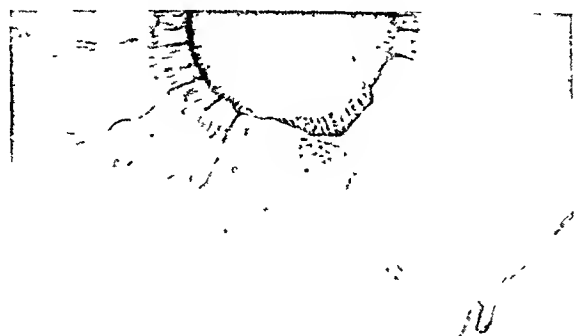


FIG. 341. Leiomyoma of the iris. (After Frost.)

margin to a little more than one-half the width of the iris. The mass was wedge-shaped with its apex directed toward the pupillary margin, and extended from 4 to 6 o'clock (Fig. 341). The thickest portion was near the pupillary margin and gradually grew thinner toward the root of the iris. Small blood vessels were visible in the stroma making their way in the iris tissue from the periphery toward the mass. A dilated tortuous anterior ciliary vessel was seen in the same sector as the tumor. There were no precipitates on the posterior surface of the cornea, and no evidences of active inflammation were present. Histologically, it was a vascular tumor composed of interlacing bundles of closely packed elongated spindle-shaped cells with considerable granular eosinophilic cytoplasm and long oval nuclei. The tumor is of greater thickness at the level of the sphincter irides, with which it appears to be continuous. Differential staining with the gold impregnation method also revealed the presence of the characteristic myoglia fibrils and a diagnosis of leiomyoma of the iris was made."

ANGIOMAS

Several cases of hemangioma of the iris have been reported in the literature. According to Duke-Elder only the one reported by Rodin (1929) was proved histologically. Angiomas have been described as

highly vascularized circumscribed tumors that protrude from the iris surface and that tend to bleed easily. De Wecker described a case in which a pea-sized angiomatous growth resembled a blackberry in appearance.

The occurrence of such growths in connection with Sturge-Weber's syndrome (nevus flammeus) of the face and with glaucoma or buphthalmos has been noted on several occasions (Knapp, 1928; Tyson, 1932; Evans, 1937⁴²⁵). In the case of a young woman who had nevus flammeus, glaucoma, and engorged conjunctival vessels (Vol. I, page 249), several large tortuous vessels were seen in the iris. This was also noted by Evans⁴²⁵ in a 12-year-old boy in whom nevus flammeus was associated with hemangioma of the choroid. In Daily's case, a 5-month-old infant, there was a very narrow dark-brown growth in the angle of the anterior chamber. There was considerable blood in the anterior chamber, and the eye was hard. Histologic examination revealed a hemangioma of the ciliary body, which projected into the anterior chamber at the infiltration angle for about 2 mm. This portion was necrotic and most likely was the source of the hemorrhage. Vascularized growth masses in the iris simulating angiomas have to be differentiated from the granulomas and from malignant growths, especially the so-called "leukosarcomas." This requires a biopsy in most instances. In this connection, the so-called vascularized connective tissue growth — nevus vasculosus — should be mentioned. This type of growth tends to develop in the neighborhood of the sphincter. Fuchs (1913)⁴³⁷ described it as a vascular tumor of congenital origin. They may remain silent for many years or at times begin to grow. Active growth of such a lesion would indicate that it represents a new growth of the vessels themselves.

MALIGNANT MELANOMA OF THE IRIS

Today benign melanoma and malignant melanoma as well (following the classic ideas of Masson, and with specific reference to the eye, those of Theobald) are considered to be neuro-ectodermal in origin, deriving from the cells of Schwann. The reader is referred to the original papers on this subject. Actually primary malignant

melanoma in the iris is very rare. In most cases histologic evidence will reveal forward extension into the iris from tumors in the ciliary body. Although a considerable number of cases have been reported in the literature in which malignant melanomas have been observed to arise from previously seen pigmented spots or freckles (Reese), it is not at all certain that this is so. For instance, Fuchs (1917) thought that pigment spots previously seen at the site of a malignant melanoma were not physiologic iris pigment spots or nevi but actually the early evidence of an already slow-growing malignant melanoma. In addition there is the added fact of the rarity of malignant melanoma as compared to the universality of the so-called "physiologic pigmented iris spots." On the other hand, the tendency to malignant growths in eyes having melanosis oculi has been noted many times. It would seem that eyes that are abnormally pigmented congenitally are more predisposed to develop malignant melanomas. This is also the case in persons with skin nevi. In other words, those in whom a tendency to congenital pigment exists are more likely to develop malignant melanomas than others. However, the tumor does not necessarily develop at the site where the congenital pigmentation is most marked. This point was brought out by Van der Hoeve's observation (1924) in which there were numerous black spots in the fundus of a right eye of a man, who in his left eye had a malignant melanoma removed by iridectomy. Recently, Reese * has shown the frequent occurrence of benign melanomas (pigment freckles) of the iris in microscopic sections of eyes harboring malignant melanomas of the uveal tract.

* Reese states: "The histologic difference between the melanomas of the iris seen in normal eyes and those seen in eyes with malignant melanoma seems to be one of degree. In the case of the normal eye the lesion is a localized accumulation of densely pigmented melanoblasts only a few cells thick along the anterior surface of the iris. In fact the lesion could be characterized as merely a localized thickening of the anterior limiting layer of the iris, which in some instances has the appearance of being proliferated endothelium. Such a lesion may sometimes contain little or no pigment (similar to Wofrűn bodies [author]). The melanomas of the iris in eyes with malignant melanomas are more extensive lesions. They tend to be slightly more elevated above the iris surface and to extend deeper into the iris stroma." Considering the nature of the multiple manifestations of melanomas, he gives consideration to the idea that perhaps some cancerigenic agent acts on the whole uveal tract causing a malignant melanoma at one site and more or less benign melanomas at other sites. This is in line with what occurs elsewhere in the body, notably in the Phakamatoses (von Recklinghausen's disease).

Biomicroscopically, it has not been possible at an early stage to differentiate between a simple physiologic iris freckle (benign melanoma) and a malignant growth, although differences (degrees of maturity) in the character of the cells must exist. Nor is it always possible in the early state to recognize the transformation from the benign to the malignant status, especially when development is slow. It has been suggested that the appearance of vessels in the iris or even at the limbus in the sector occupied by the lesion is a significant indication of such a change. In several cases of malignant melanoma, which came under my observation, I could make out no increase in vascularity in the iris, even after the mass attained considerable size. However, secondary effects (toxic and pressure) may cause the appearance of even very large iris vessels in the neighborhood of a tumor. This is usually a manifestation of a late phase, as a criteria of malignancy. Vascularity of the lesion itself is better manifested in leukosarcoma and in carcinoma. Undoubtedly the *sine qua non* in this differentiation is visible signs of progressive growth. Repeated biomicroscopic examination with accurate measurements (using the ocular micrometer) is of greatest importance in detecting changes in shape, volume, or extension, all characteristic of growth. Enlarged anterior segment kodachrome photographs are valuable in recording the actual appearance and changes.

It has been long recognized that malignant melanoma of the iris may occur in young individuals, and even at an earlier age than in the choroid. The growth, as previously mentioned, may start as a dark brown to blackish spot anywhere on the iris surface, indistinguishable from an ordinary physiologic freckle. Gradually and quietly over a period of months, years, or decades it increases in size and thickness, eventually projecting from the iris surface to form a well-defined nodule. Its surface soon tends to become bossellated and corrugated (Plate LVII). However, I have seen cases in which the surface remained smooth and velvety. In these the tumor invaded the iris from the ciliary body either anteriorly at the chamber angle or appeared in the pupillary area after extending axially just behind the iris in the posterior chamber. Frequently it happens that small

freckles are seen in the neighborhood or elsewhere on the iris surface. If these were not previously present, i.e., before the appearance of a tumor, they might represent daughter growths, either multiple origins, implantations, or direct extensions. Only close observation over long periods of time with the biomicroscope to determine active growth will answer the question as to whether these freckles are benign (congenital) or are part of the malignant process. In any event the appearance of new freckles on the iris in the vicinity of a tumor should be looked on with suspicion. This holds true not only for well-formed flat freckles but even for pigment dust, dispersed over the iris surface. This is demonstrated by a case of Gilbert's in which a distinct difference in color and regularity of the iris surface occurred as compared to that of the unaffected eye. Pressburger (1932) reported on the character of tumor cells dispersed on the iris and lens capsule. With the biomicroscope he saw "sarcoma" cells on the anterior lens capsule. They appeared in the form of fine lines and starlike, branching, gray to brownish spots. Characteristically these cells had long processes (outriders) which he believed differentiates them from the well-known small starlike cells seen in congenital remains of pupillary membranes. In every case, particularly if the tumor is seen in the peripheral parts of the iris, it is important to know whether the tumor is primary to the iris or whether it is an extension from the ciliary body. This question is very important (though frequently insoluble) when the tumor seems to arise from the iris root since one originating in and localized to the iris might conceivably be removed by iridectomy whereas extension into or from the ciliary body demands an immediate enucleation. Unfortunately, after the tumor in the region is raised above the iris surface, it usually is not possible to inspect the chamber angle just back of it. The wider the base of the growth is at the iris root the more likelihood there is that it originates from or involves the ciliary body. This is especially true with the rarer flat or ring type of malignant melanoma. As in the case of the ciliary body, the natural circular architecture of the iris tissue at the lesser circle and at the iris root favors ringlike or circular extension. Such an infiltration may occur

within the stroma so that very little thickening or elevation above the iris surface is seen. But as a rule clinically the iris assumes a peculiar dirty brown to black discoloration. Adjacent parts of the stroma may undergo varying degrees of atrophy and vitiligo; as a result the iris assumes a grossly mottled appearance in which darkly colored tumorous areas are sharply contrasted with depigmented areas. Occasionally it has happened that histologic examination reveals that the entire iris stroma has been replaced by such a form of growth. Although the site of the origin of malignant melanomas of the iris is probably the anterior border layer deeper involvement within the posterior pigmented retinal layers can also occur.

Several cases reported by various authors has demonstrated the fact that extension of tumor growth from peripheral parts of the iris to the ciliary body may occur. Reese⁵⁷³ has reported such a case. Malignant melanomas arising at the lesser circle or in the pupillary zone tend to grow toward the pupillary margin and generally reach this area long before they involve the ciliary zone and chamber angle. Extension into the pupillary zone causes pupillary distortion and even visual disturbances if part of the mass encroaches on the pupillary area.

No two tumors grow alike or at the same rate of speed, and consequently it is not always possible to classify the stages of growth. Anterior chamber hemorrhage may occur at any period. An initial quiet period may be followed by increased intra-ocular pressure with inflammatory symptoms, complicated cataract, and eventually blindness.* Extra-ocular perforating extensions may or may not occur. In one case this was the first manifestation of a malignant melanoma of the ciliary body. Two small black spots, about 0.5 mm. in diameter, appeared below in the sclera in the vicinity of one of the dilated episcleral veins. Two years later a small nodule was seen on the iris near its root on the angle of the anterior chamber in the same sector (below). Six months later, with the pupil widely dilated, a large

* It is conceivable that with marked degeneration and shrinkage of the eye and as a result of spontaneous necrosis from cutting off the blood supply, choroidal tumors may be found accidentally in shrunken globes.

bulging mass extending toward the vitreous could be seen below extending from the ciliary body (Plate LVII, fig. 1).

LEUKOSARCOMAS

This type of nonpigmented iris tumor is rare. It is easily recognized by its yellowish-white color and high degree of vascularization (Plate LVII, figs. 2, 3). The vessels may stand out as thick, widened cords and hemorrhages in the anterior chamber are common. The tumor may start out as a small nodule, but after slow progressive growth it may infiltrate the entire iris. Frequently forward extension occurs from a primary tumor of the ciliary body into the chamber angle and iris. As with melanotic tumors, involvement of the ciliary body may be followed by extra-ocular extension early in the overlying scleral area by means of the emissaria. In the same way tumors involving the chamber angle cause interference with the drainage channels and secondary glaucoma may rapidly supervene. Zentmayer⁶⁸⁵ reported a case in which a small mass, arising in the pupillary zone, encroached over the pupillary border. It was entirely removed by iridectomy.

METASTATIC CARCINOMA OF THE IRIS

Metastatic carcinoma of the iris occurs much more rarely than in the posterior parts of the choroid. According to Sanders⁵⁹⁶ only 4 cases are on record in which the growth has been shown histologically to be limited to the iris itself. In cases reported by Toulant⁶³⁸ in 1916 (not examined histologically) both irides were involved. In 2 out of these 4 cases the primary lesion was in the breast.*

In the iris as elsewhere in the uvea, the histologic structure of the growth duplicates that of the primary lesion (mucous or glandular)

* According to Lemoine and McLeod:⁵¹⁹ "Carcinoma of the breast tends toward hematogenous metastasis, releasing comparatively large numbers of cells into the circulation. A certain proportion of these cells find their way into the ophthalmic artery and are distributed to the tissues supplied by it. Those which reach the choroid through the posterior ciliary arteries and, to a less degree, those which reach the iris and ciliary body through the anterior ciliary arteries find nutritional conditions suitable to grow, *elsewhere* they perish. The difficulty of entrance into the ophthalmic artery (this vessel arises from the internal carotid artery at a right angle [M. L. B.]) may partly account for the low frequency of ocular metastasis. In other words, we are postulating a specific affinity of uveal tissue, particularly the choroid, to cells from carcinoma of the breast."

PLATE LVII

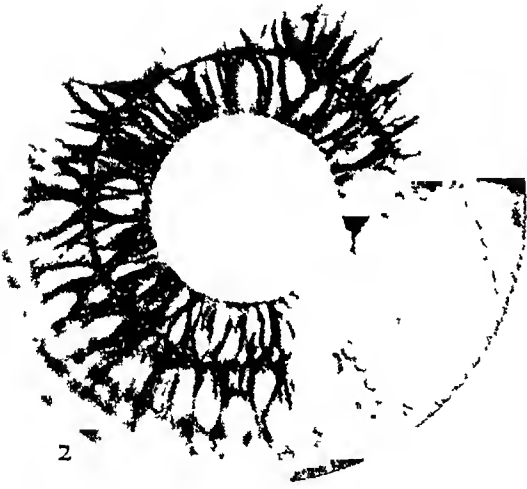
FIG. 1. Malignant melanoma (melanosarcoma) of the iris with deformed pupil.

FIG. 2. Leukosarcoma. Diffuse illumination.

FIG. 3. Leukosarcoma. Same case as Figure 2. Direct focal illumination.

FIG. 4. Malignant melanoma of the iris (deformed pupil).

FIG. 5. Malignant melanoma of the ciliary body extending into the iris, above.
Detached particle, below.



and clinically usually appears as small well-defined nodular to massive swellings, varying from yellow to grayish brown in color located anywhere or in any zone of the stroma. These lesions are well vascularized, the vessels varying in size. Sanders' case occurred in a woman who six years previously had a hysterectomy. Below at the base (root) of the iris there was a flesh colored mass about the size of a pea having a few dilated vessels on its surface. There were no signs of active inflammation. The pupil was dilated and fixed, and the intra-ocular pressure was elevated. The fundus was normal. Within two months the tumor grew rapidly until it almost filled the anterior chamber. A grayish infiltration was noted deep in the lower half of the cornea. Eventually the invaded cornea ruptured. The mass shrank to below the pupil and the perforation became closed by granulation tissue. Postmortem examination revealed that the iris stroma was completely replaced by the tumor mass.

Although the growth is slow in the early stages, affecting mobility over the involved area, it soon tends to advance rapidly invading the chamber angle; this may lead to increased intra-ocular pressure. Finally the mass may partly or entirely fill the anterior chamber, eventually invading the cornea. As a consequence of necrosis, a secondary iritis (although infrequent) may occur. If this should occur early when the nodule is small, confusion in diagnosis with a granulomatous lesion is conceivable, but the tendency to rapid growth and the history or suspicion of organic carcinoma elsewhere (especially the usual history of breast removal in women) will support the diagnosis of a carcinoma.

Chapter Twenty-Three

THE NORMAL LENS; BIOMICROSCOPIC APPEARANCE

THE LENS

GENERAL Considerations. Biomicroscopic examination of no other part of the eye has been so fruitful and so rewarding as that of the lens. The use of the focal beam (especially the optic section) in the living eye has opened the door to new vistas in the development, morphology, and pathology of the lens, comparable to the advances following the discovery of the ophthalmoscope. Before the advent of biomicroscopy our concept of the living lens was inadequate as it was based for the most part only on what could be seen with the ophthalmoscope. Histologically, it was only by the employment of special staining methods (Rabl) that its fiber structure and suture systems became apparent. Ordinary methods of fixation and staining provide very little if any information concerning the actual structure, normal or pathologic, of the lens. The fixing and hardening histologic processes frequently make it impossible to cut the lens properly. With the ordinary hematoxylin and eosin staining the lens appears uniformly pink and it is often impossible to differentiate the normal morphology from the pathologic. Post-mortem changes result in disintegration and vacuolization. The normal concentric (internal) zones of discontinuity and the suture systems, delicate changes like lamellary separations, water slits, vacuoles, opacities in the cortex, opacities of the inner nuclei, so beautifully seen biomicroscopically, are all unrecognizable as such in histologic preparations. However, it is possible to discern some of these delicate changes in the freshly removed lens. Such material is difficult to obtain and must be examined within a half hour after

death since vacuoles and degeneration of the fibers occur almost immediately afterward.*

The normal lens continues to grow throughout life; biomicroscopically it presents different pictures with each epoch. The focused beam not only allows recognition of the developmental changes but topographically permits of their exact location. This holds true for pathologic and traumatic lesions as well. The lens consists of epithelial tissue only (except for its cuticular capsule) and grows by the transformation of its epithelial cells into fibers. The older fibers recede into the depths and become "hard" as the newer, soft fibers develop and grow around them. The progressive growth of the lens fibers from the earliest embryologic tissue results in a stratification of the internal architecture which ultimately forms the so-called zones of discontinuity (Fig. 350; Plate LVIII, figs. 1, 2). These are zones in which there are sharp changes in indices of refraction, manifested biomicroscopically by stripes or bands of increased relucency. In optics such bands or stripes are known as zones of discontinuity (Vol. I, page 76; fig. 70). In addition to these stripes definite systems of suture figures are seen. After the earliest embryonal stage the lens fibers grow from the equator toward the axial region of the lens. The meeting place of the fiber ends form the sutures. As will be shown later, this complex internal architecture of the growing lens seems not only to insure its development and maintenance, as an optical instrument of proper shape (its only purpose) but also to allow for the processes of accommodation. Before the advent of the biomicroscope, only the lens nucleus, the surrounding cortex, and the capsule were differentiated. The Sanson-Purkinje reflex hinted at the presence of internal reflecting zones, but the exact internal architecture of the lens was unknown until after the invention of the biomicroscope.

For the purposes of orientation, the embryology of the lens will be reviewed briefly. Contrary to what occurs in other "epidermoidal"

* According to Vogt, if lenses are fixed *in situ* by the mercury bichloride and platinum chloride method of Rabl, it is possible to examine fibers and their changes. However, even with this method one has to be careful in evaluating the findings because of secondary changes due to imbibitions and beadings.

PLATE LVIII

FIG. 1. Optic section through the normal lens. Anterior part is in focus; posterior parts seen in divergent (unfocused) rays. Observe zones of discontinuity, and the shagreens (specular reflex) of the capsule.

FIG. 2. Optic section through the normal lens. Posterior parts in focus, showing the band of the posterior adult nucleus, shagreen of the posterior lens capsule, remains of the hyaloid vessels, the retro-lental space (hyaloid vitreous). Anterior parts of lens now out of focus. Note the shadow from the small congenital opacity in the embryonal nucleus.

FIG. 3. Shagreen (specular reflex) of the anterior capsule and the anterior adult nuclear band. High power.

FIG. 4. Shagreen (specular reflex) of the posterior capsule and the posterior adult nucleus. High power.

FIG. 5. Direct focal illumination. Showing composite view of the beam passing through the lens of a young man aged 17. Wide beam.

FIG. 6. Direct focal illumination. Narrow beam (optic section) through the lens shown in Figure 5.



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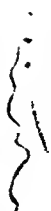
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tissues (e.g., skin, nails, hair, etc.), the lens capsule prevents any shedding or casting off of early or "dead" tissue and hence throughout life it is possible to see even the parts which embryologically were present from the very beginning. After birth the rapidity of growth diminishes, the central or older parts become compressed or sclerosed as new or younger fibers grow around them. That the growth of new fibers is continuous during life was proved clinically by Vogt. In the case of the disseminated whitish cataractous spots seen subcapsularly after an acute attack of glaucoma it was noticed that after months or years these circumscribed spots were pushed into the deeper cortical layers. New clear fibers were interposed between these white spots and the capsule. In the "congenital" polar cataracts, separation of the opacity by clear fibers resulting in the formation of an imprint or duplication opacity is another instance of progressive growth of the outer part of the lens.

As early as the second week of embryonic life * there is a slight elongation or thickening of the cells (lens plate) of the surface ectoderm where it lies in contact with the optic vesicle (Fig. 342). A depression of surface ectoderm of the lens plate soon follows and forms the lens pit. The pit deepens and the cells at the opening of the pit come together to form a stalk and a lens vesicle. During the fourth week (10-mm. stage) the vesicle separates from the surface ectoderm and appears as a hollow globe or cyst, its walls being composed of a single layer of surface ectodermal cells. Anteriorly these cells become the subcapsular epithelium. The cells forming the posterior wall of the vesicle now begin to elongate (primary lens fibers) and grow forward until they fill the cavity of the vesicle. This process occurs during the fifth and sixth week, the embryo increasing in size to about 20 mm. at the end of the sixth week. At about the 13-mm. stage a hyaline capsule is secreted. This comprises the posterior portion of the hyaline lens capsule. The anterior parts

* According to the time of development of the lens nuclei before birth, I prefer to call the completed primary lens vesicle the "embryonic" one. Especially since the period of its growth occurs during the first three months of life when the developing individual is known as an embryo. For the same reason, the nucleus which starts to develop after the completion of this stage (i.e., after the third month or at the time when the developing individual becomes a fetus) is the "fetal nucleus."

of the hyaline capsule are probably developed later, and peripherally some of the layers are derived from the lenticular expansions of the zonule, consequently the capsule is best developed in this region.

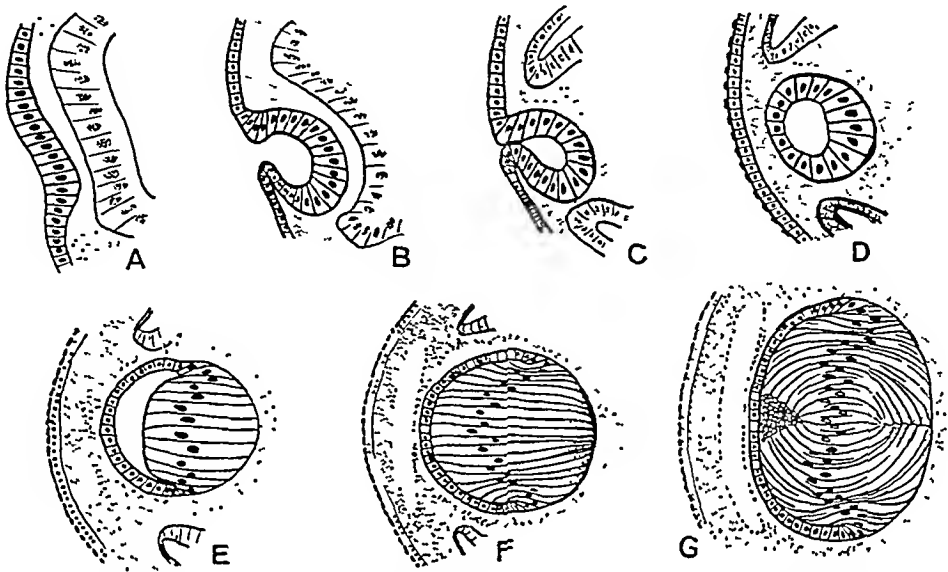


FIG. 342. Embryonal development of the lens. Stages in the development of the human lens. A, the lens plate; B, the lens pit; C, the lens pit closing; D, the lens vesicle; E, primary lens fibers complete; F, secondary lens fibers and sutures (After Mann)

Clinically, external portions of the hyaline capsule are seen in cases of exfoliation of the lens capsule (senile, glaucoma capsulare, heat cataract).^{*} The fact that the cells lining the posterior wall of the vesicle elongate explains the absence of epithelium on the posterior capsule. In extracapsular cataract operations the remaining thin transparent posterior hyaline capsule ordinarily does not interfere with vision but is easily visible biomicroscopically.

In the fully formed lens the elongated primary lens fibers that filled the primary vesicle are recognized biomicroscopically as a dark almost nonrelucant area in the center of the lens (the central dark interval), the cellular outline of these cells no longer being apparent.

After this stage and during the rest of fetal life as well as the remainder of the life of the individual, new fibers (secondary fibers) are continuously derived from the cells of the anterior capsular

^{*} Employing special methods, Kolliker (1854),⁵⁰⁵ Ivanoff,⁴⁸⁷ Berger,³⁶⁵ and Schirmer,⁶⁰⁰ and Vogt (1914)⁶⁴⁶ demonstrated that the hyaline lens capsule consisted of numerous lamellae. This was revealed by teased or macerated preparations or by the use of chemicals, acids and alkalies, salts and trypsin. Vogt was able to demonstrate ten layers.

epithelium in the region of the equator. After the completion of the growth of the primary lens vesicle or embryonal nucleus, these fibers extend from the equator and grow around the primary ones

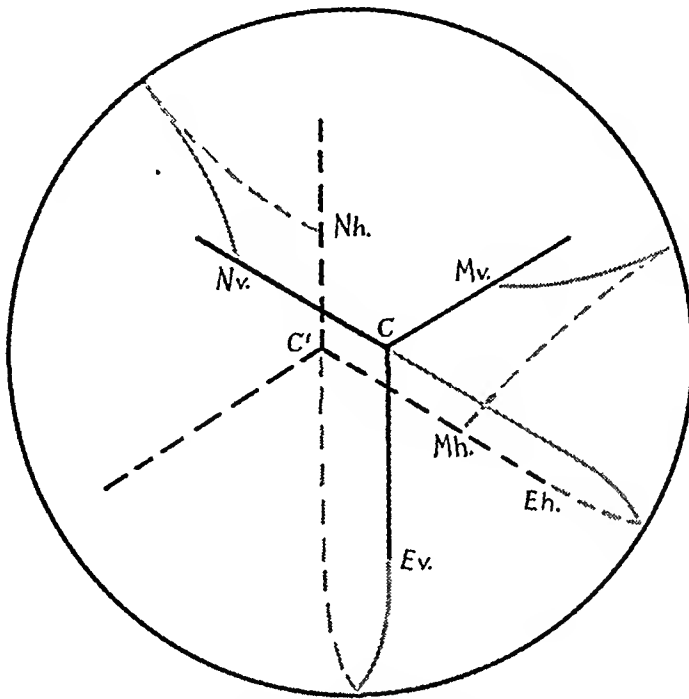


FIG. 343. Diagram of the course of the lens fibers (human newborn). The erect anterior Y-suture is represented by solid black lines. The fibers are represented by grey lines, solid for the anterior zone and dotted for the posterior ones. All the fibers are of equal length. A fiber which originates from the posterior suture arm, *Eh*, ends at the center of the Y-suture, *C*. A fiber which originates from the middle of the anterior suture arm, *Mv*, ends at the middle of the posterior suture arm, *Mb*. A fiber originating from the outer third of the anterior suture arm, *Nv*, ends at the inner third of the posterior arm, *Nh*, etc. (After Vogt.)

from all directions to form the two coffee-bean-shaped nuclei which after birth are located at a slight distance from the capsule. Between these nuclei is the dark interval (true embryonal nucleus). The new nucleus contains the well known Y-sutures (recognizable at the 35-mm. stage [8 to 9 weeks]) and is known as the inner embryonal (Vogt) or better as the fetal nucleus (Koby, Mawas and others). Its inner surfaces face the dark interval. The sutures, anteriorly an erect Y, posteriorly an inverted Y, are really potential spaces resulting from the line of contact or abutment of the fiber ends. (See Vol. I, Fig. 70; Figs. 343, 344 A and B, 365, 366.) It is in these simple fetal sutures that the relations between the suture formation and fiber growth are best seen since in the more complicated many

branched sutures (cortex and adult nucleus) it is not possible to trace the course of the fibers so easily. A fiber growing short (to form the extremity of one of the limits of the Y) anteriorly will

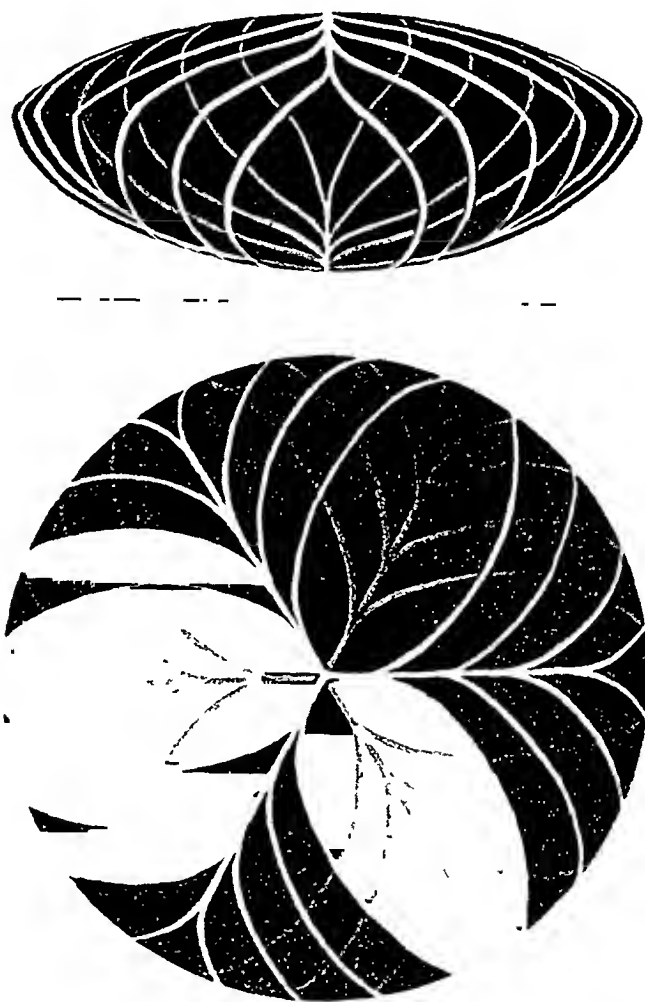


FIG. 344. Plastic representation of fiber growth in the formation of sutures.

grow longer posteriorly to end in the center of the suture (Fig. 343). A fiber growing to a point midway in the suture arm anteriorly will end correspondingly at the midpoint of the suture arm posteriorly. In this way it will be seen that fibers in one concentric zone are of the same length. This type of growth anteriorly and posteriorly insures the spheric development of the lens (Fig. 344). If all the fibers grew equally to meet in the center of the lens a discoid concavity would result at this place and the lens would have a to-

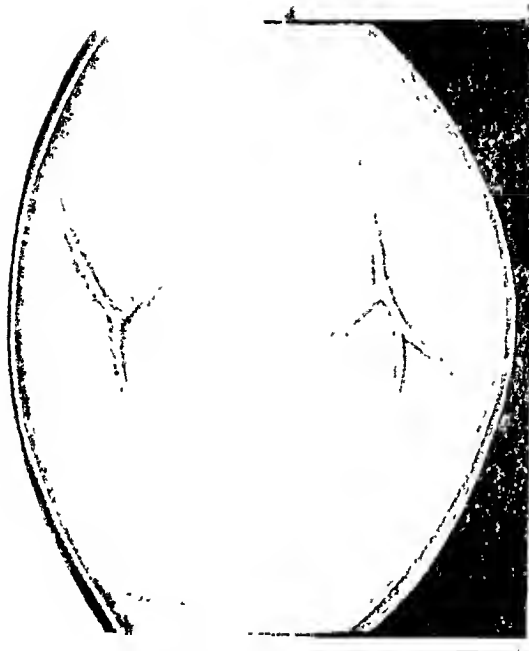


FIG. 345. Direct focal illumination of the lens of a 4-months-old baby. The sutures are found a little distance behind the capsule stripes.

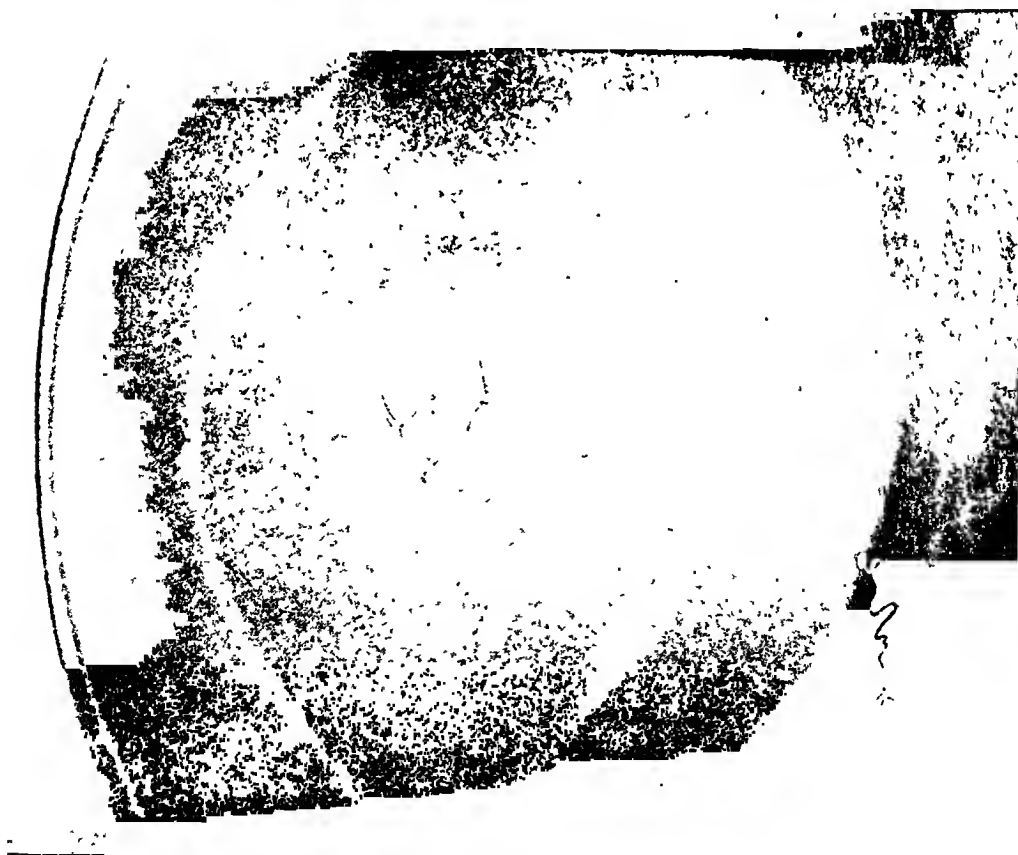


FIG. 346. Direct focal illumination of the lens of an 11-year-old child.

roidal shape; and hence would lose value as an optical instrument. At birth the Y-sutures are still close to the capsule (Fig. 345), but are already separated from it by a thin layer, the developing adolescent nucleus. From this point on during the remainder of the individual's life it is this part of the lens in which development and growth of new fibers occurs. In young adults it already is possible to see the division in this part of the lens (anteriorly and posteriorly) into three main parts: (1) the adolescent nucleus; (2) the so-called "adult" nucleus; (3) the cortex, between the adult nucleus and the capsule (Fig. 346). In optic sections, lines of separation or disjunction are seen, i.e., lines immediately adjoining the capsule stripes. These lines (anterior and posterior) which are found even in the youngest individuals are closest to the capsule stripes in the polar regions but tend to diverge slightly (as do the other stripes) as they extend toward the equator. The importance of zonular traction in governing and maintaining the flatness of the lens postnatally is seen in the cases (microphakia and spherophakia, page 1341) in which aplasia of the zonular fibers occurs. In these cases lack of zonular traction results in a ball-like (spheric) lens. The action of the zonula may play a great role in length and branchings of the extrafetal sutures during life or at any rate until the time of presbyopia. With increase of lengthening and branching of the sutures (Vogt) the fibers that abut them have to become wider (they do this by becoming less thick) in axial regions in order to compensate for the greater area they have to occupy as compared with the region of their origin in the periphery. Likewise zonular traction and site of insertion of the zonular fibers may explain the lesser curvature (flattening of the anterior surface, which is a normal finding. The greater tension anteriorly (and hence the greater flattening) may be due to the more axialward insertion of the zonular fibers on this surface and to the difference in traction between them and the posterior zonular fibers.

Blood Supply. In fetal life — for the purpose of nourishment — a vascular net derived from branches of the hyaloid vascular system in the vitreous forms on the posterior capsule (posterior vascular cap-

sule). From this net parallel vessels (capsulopupillary portion)* run anteriorly around the lens equator or between it and the margin of the optic cup. These vessels anastomose with the annular vessel (cho-

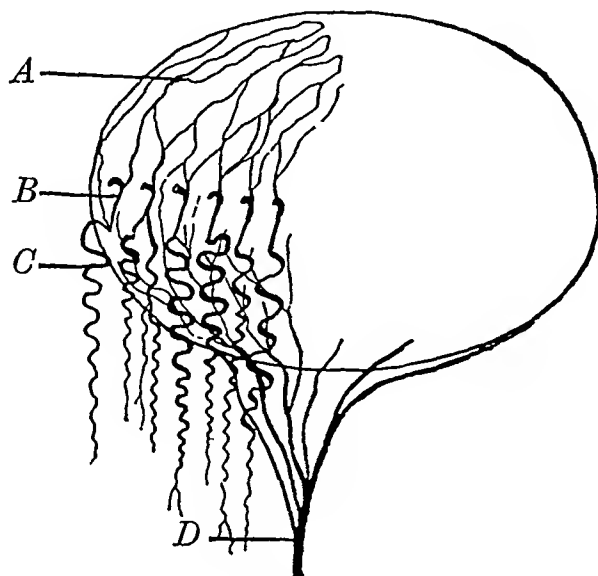


FIG. 347. Diagram of vascular capsule of the lens. *A*, anterior part (pupillary membrane); *B*, Capsulopupillary part; *C*, Vasa hyaloidea propria and posterior vascular capsule; *D*, Hyaloid artery. (After Mann.)

roidal veins). Both are seen by the 16-mm. stage and reach their maximum development by the 40- or 50-mm. stage when regression occurs. At about the 28-mm. stage branches from the capsulopupillary vessels and from the long ciliary vessels form a series of loops which cover the anterior capsule of the lens (Fig. 347). These form the anterior vascular capsule and the pupillary membrane. (See chapter on Iris, page 755.) At the 48-mm. stage the more developed peripheral parts lose their connections with the capsulopupillary vessels at the time the ectodermal part of the iris begins to extend forward. By the seventh month marked regression and atrophy of the posterior, lateral (capsulopupillary) vessels has occurred and beginning atrophy of the anterior vascular capsule is noted. However, the arcades of the central portion (pupillary membrane) do not disappear until almost term. The peripheral system which loses its connection with

* According to Terry, the term "lateral or capsulopupillary" for a portion of the tunica vasculosa lentis is confusing. He suggested the term "intermediary portion" of the tunica vasculosa lentis as more suitable.

the capsulopupillary vessel persists to form the deeper vessels vascularizing the iris stroma. Although grossly there appears to be a complete atrophy of all the vascular capsule of the lens the biomicroscope has revealed that this is not so. In every eye vestiges of the hyaloid system (*tunica vasculosa lentis*) are observed not only on the posterior lens capsule but also in the anterior parts (hyaloid or retro-lental) of the vitreous. This is demonstrated by the universal presence of the small hyaloid remains (corkscrew remnant), the arcuate line and occasionally the presence of nodular circumscribed whitish bodies, the so-called spurious posterior polar cataracts, which Vogt designated as "*corpusculum nodularis hyaloidea*," on the posterior lens capsule (Figs. 373, 374). Anteriorly, in about 25 per cent of normal people (according to Mann) there are tags of the pupillary membrane attached to the lesser circle of the iris. Not infrequently a central attachment to the lens capsule occurs. A large number of lenticular anomalies may be due to or connected with the persistence of pupillary membranes. Not uncommonly, we find pigmented or whitish starlike remains, threads (white or pigmented), or small white thickenings on the anterior capsule (Vol. I, Plate XXXVIII). In addition (with dilated pupil) fine radial lines or stripes are seen occasionally peripherally on the anterior capsule. These are known as retro-iridal lines or stripes, and they probably represent remains of the capsulopupillary vascular membranes or, as Vogt prefers to call them, the "*tunica retro-iridalis*" since in the histologic sense they are not true membranes.

TECHNIQUE OF BIOMICROSCOPY OF THE LENS

With the obvious exception of sclerotic scatter, all the methods of illumination applicable to the cornea are employed in the examination of the lens. (See Vol. I, Chapter Three.) These are:

1. Diffuse illumination
2. Direct focal illumination (wide and narrow beam)
3. Specular reflection
4. Retro-illumination

5. Indirect illumination
6. Diapupillary transillumination of the lens in fundal reflex (transillumination of the lens by means of light reflected from the fundus)

With the undilated pupil only the axial regions of the lens are accessible. Unquestionably it is impossible to make an accurate survey of the lens unless the pupil is maximally dilated since a large percentage of lens changes either begin or are localized in the periphery. The examination room should be dark, and the observer dark-adapted. The light should be directed first from the temporal side and then from the nasal in order to view both medial and lateral portions of the periphery. Even with the pupil maximally dilated, e.g., as obtained by the use of a cotton pledget soaked in 1:1000 epinephrine or 10 per cent neosynephrine, it is impossible to see the actual equator or the zonule. This is only possible with coloboma irides or subluxated lens, or when a condition such as spherophakia is present. However, by directing the patient's gaze upward, downward, and sideways it is feasible to examine most of the lens.

DIFFUSE (AFOCAL) LIGHT

When one employs the pre- or postfocal part of the beam (Vol. I, page 69) for illumination of the lens it is possible with sharp focusing of the microscope to see certain surface areas in their entirety (Fig. 348). For example, the anterior lens surface with part of its shagreen, the surface and suture system of the adult nucleus (only in older individuals), the entire embryonic Y-sutures and the posterior capsule (especially when altered pathologically). This method corresponds to the ordinary method of oblique examination with the loupe and lens except for the fact that observation of details is aided by the use of the binocular microscope. Using a condensing lens and loupe Tscherning⁶⁴² noted the now well known bright "image" within the pupil. When the light is bright the pupil appears white (in a normal lens) as if a mature cataract was present. This image results partly from specular reflection (shagreen) of the cap-

sule and surface of the adult nucleus. With the loupe especially in older persons the suture system of the adult nucleus can be made out. Using the unfocused part of the beam together with the microscope

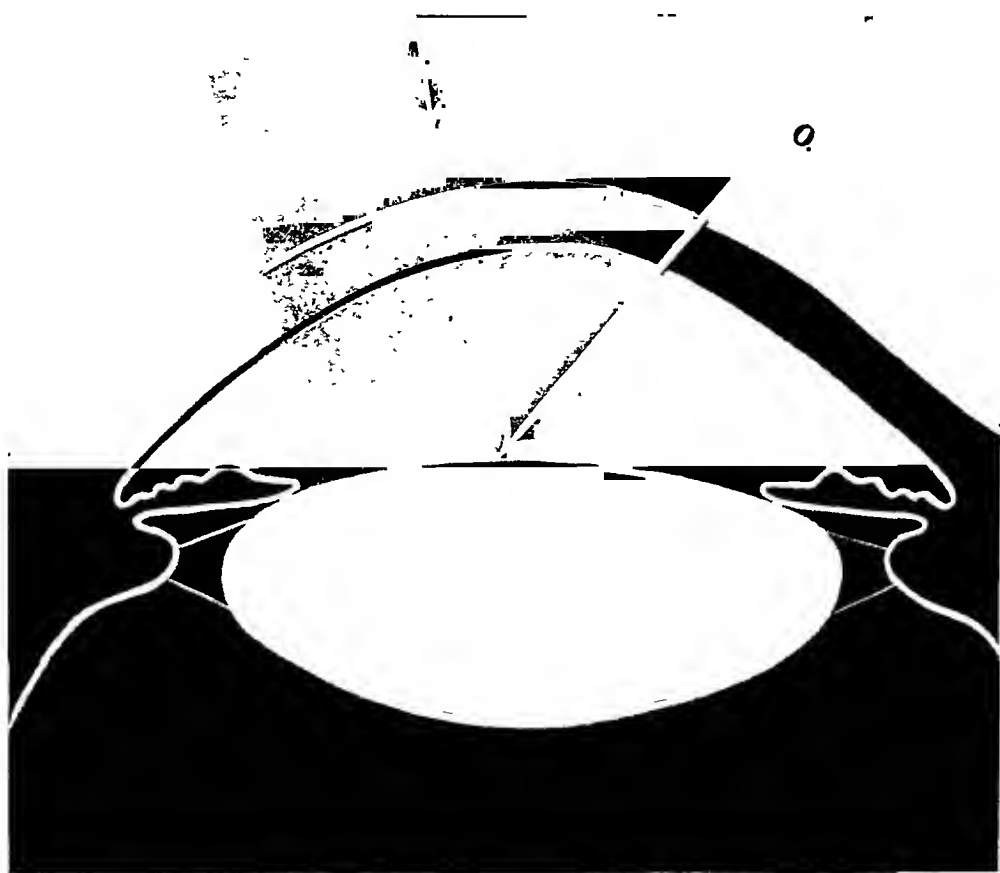


FIG. 348. Diagram illustrating technique of diffuse illumination.

such details can be seen more distinctly. The entire extent of the posterior Y-suture and certain types of localized lens changes, such as congenital cataracts, polar cataracts, lenticonus, saucer cataracts, can readily be seen "en pleine." However, this method does not allow for accurate localization except as one judges it by focusing the microscope, using known fixed points of reference, e.g., the anterior or posterior lens capsule.

DIRECT FOCAL ILLUMINATION

If the focused beam is allowed to pass diagonally into the pupillary opening it will be seen, that similar to the appearance in the cornea (but unlike the anterior chamber which normally appears dark) a

circumscribed gray relucient block is formed (Vol. I, Fig. 70). The unilluminated and surrounding portions of the lens are relatively dark. The transparent tissues of the eye are actually gels and hence



FIG. 349. Diagram illustrating direct focal illumination. Anterior parts of the lens in the focal part of the beam. Posterior parts of the lens in the post focal of the beam. The prefocal part of the beam is located in the cornea.

will show the Tyndall phenomenon in varying degrees (Vol. I, page 72), depending not only on the intensity of the light but also on tissue density. As the light passes through the cornea and forms the parallelepiped, a proportional amount of the light's intensity is "used up" and this in itself tends partly to weaken the apparent relucency of the lens block. The illuminated lens segment or "phantom," which optically represents a section through the lens thickness, is the visible manifestation of its optical properties (Fig. 349). In summation, these are refraction, reflection from the surfaces (zones of discontinuity) of its layers, internal dispersion, polarization and fluorescence. At each stratum in the lens where a difference

in index of refraction abruptly occurs we become aware of a zone or band of increased brightness. As the light passes through the lens posteriorward it will be seen that there are about six main whitish

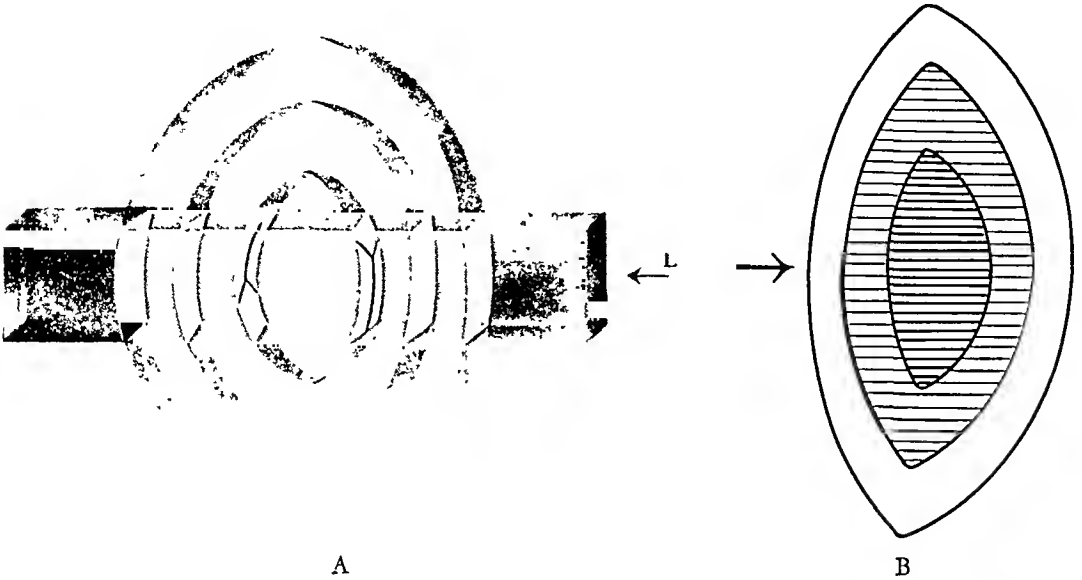


FIG. 350. A. Diagram showing passage of the beam L from left to right through the lens and the illumination of the major zones of discontinuity. These are, in order, 1, capsule stripe; 2, surface of the adult nucleus; 3, anterior fetal nucleus; 4, posterior fetal nucleus; 5, surface of the posterior adult nucleus; 6, the posterior capsule. The fainter adolescent stripes between the adult and fetal nuclei are not shown. B. Diagram illustrating the central fetal nuclei surrounded by the adult nuclei which in turn is surrounded by the cortex.

stripes or zones of discontinuity (Fig. 350 A, B). The width of the bands or stripes depends on the width of the beam. Between them lie darker areas. On the two central stripes the anterior and posterior fetal Y-sutures are seen. It should be pointed out that although these stripes are represented in drawings as thin layers, actually they have some thickness. Beginners frequently make the error of interpreting the zones of discontinuity as running in a sagittal direction while actually they are concentric to one another. In contrast, the sutures extend sagittally at right angles to the zones of discontinuity.

These zones of discontinuity result from and represent the internal architecture of the lens. It should be remembered that owing to the fact that the sagittal diameter of the lens is from four to five times that of the cornea, it is not possible to keep its entire thickness within the focal part of the beam at one time, nor is it possible to observe all its parts sagittally with one focus of the microscope. Also

when the focused part of the beam is situated at the anterior portion of the lens (capsule), the deeper or the posterior parts are viewed in diverging (postfocal) rays (Plate LVIII, figs. 1, 2). Consequently the beam must be refocused deeper in order to reveal details in this area. To bring out clearly the reflecting surfaces (zones of discontinuity) it is necessary to have the beam as sharply focused as possible at the exact point where these sudden differences in index of refraction occur. Hence, both the beam and the microscope must be successively refocused ever so slightly as one proceeds from the anterior surface to the posterior surface of the lens. To a large degree, with an illuminating lens of 100 mm. (the one commonly used is 70 mm.) a beam of greater length is obtained, although the focal part of the beam is less luminous. Some of this refocusing is obviated if one of the diaphragms (Poser slit-lamp equipment) is used. This gives an increase in length to the focal part of the beam and with low power one can almost visualize the entire sagittal thickness of lens (without successive refocusing).

Wide Beam. Ordinarily the width of the slit opening governs the width of the focal part of the beam; narrowing the slit opening results in a corresponding decrease in width of the focal part of the beam and a proportional decrease in its light intensity. With the slit aperture wide open the beam attains its greatest luminous intensity and forms the so-called "broad beam." (See Vol. I, Fig. 66). As this passes through the cornea it forms the "parallelepiped." When the broad beam (slit opening from 2 mm. to 1 mm.) is focused on the anterior capsule of the lens, a sort of parallelepiped forms in the anterior part of the lens—the anterior surface representing a portion of the anterior capsule of the lens, the posterior surface of the parallelepiped representing a portion of the surface of the adult nucleus, while the intervening thickness represents the anterior cortex (Fig. 397). From here on the beam (post-focal part) diverges somewhat and illuminates the deeper layers in a diffuse manner. As is the case in the cornea, finer details of tissue stratification and the ability of exact localization are lost owing to reflection and diffusion. In other words the wider the beam, the

wider the area of illumination of the discontinuity surfaces will result, and consequently interference with observation of the intervening structures will occur. Also see Plate LVIII, figs. 5, 6.

Narrow Beam. For the greatest single advance in biomicroscopy we are indebted to Vogt who showed that it is possible to narrow a beam without any loss of sharpness. Not only does the narrow beam permit us to see the finer stratification of the topography of the normal lens structure (zones of discontinuity), which was previously unsuspected, but it also facilitates exact localization of even the most minute changes. The knowledge gained by this method of illumination of the lens has revolutionized our concepts of its morphology and pathology. It is only by means of the optic section that we are able to see the normal zones of discontinuity sharply. These zones represent lenticular growth epochs; this has made it possible for us to determine the chronologic age or time of origin of opacities in many instances. For example, it is now possible at a glance to identify and to differentiate between congenital, acquired opacities, presenile and senile changes, traumatic opacities, complicated opacities in ocular disease, opacities due to exogenous toxins, etc. When studying the lens the beginner should make it a practice first to direct the moderately wide beam (1 mm.) through its substance and then to practice narrowing the slit (it can be narrowed down to from 0.5 to 0.1 mm.), while observing the changing appearances through the microscope. As the slit narrows, the brighter bands or stripes (the zones of discontinuity) become increasingly visible despite the lowered luminosity of the beam. These stripes are the optical delineations of the surfaces of the nuclei at which the index of refraction changes sharply. They are seen at birth and throughout life. Thus, as Vogt has indicated a *continuous* variation in index of refraction, previously thought to occur in the lens in early life, does not exist.

When the beam is narrowed markedly, the stripes appear as lines and the less relucient spaces between them represent sections through the sagittal thicknesses of the "nuclei" (Fig. 351). As previously mentioned, with the pupil dilated to the maximum, the narrow beam

should be directed into the lens from the nasal side as well as from the temporal in order to traverse as much lenticular substance as possible, always bearing in mind the necessity of sharp refocusing



FIG. 351. Projection of the narrow beam (optic section) through the lens, showing zones of discontinuity (1-12). (Diagrammatic.)

of both beam and microscope as deeper portions of the lens are scrutinized.

One first focuses the microscope at the point of impact (anterior lens capsule), where the beam strikes the lens; then both microscope and beam are gradually and concomitantly focused deeper, always so that the focal part of the beam and the focus of the microscope fall at the same point in the lens. This process is continued until the entire thickness of the lens is traversed and the posterior capsule comes finally into view. When the anterior capsule is in sharp focus, it will be seen that the deeper portions of the lens are traversed by divergent rays, and consequently the height of the lens block is greater posteriorly than anteriorly. When the beam is sharply focused on the posterior capsule, the converse is true. Behind the posterior capsule the gossamer structure of the vitreous is seen. After the anterior and posterior capsules have each been brought into focus, then one begins to differentiate the internal zones of discontinuity and their markings (sutures). By shifting the beam slowly from side to side and by changing the direction of the illumination from

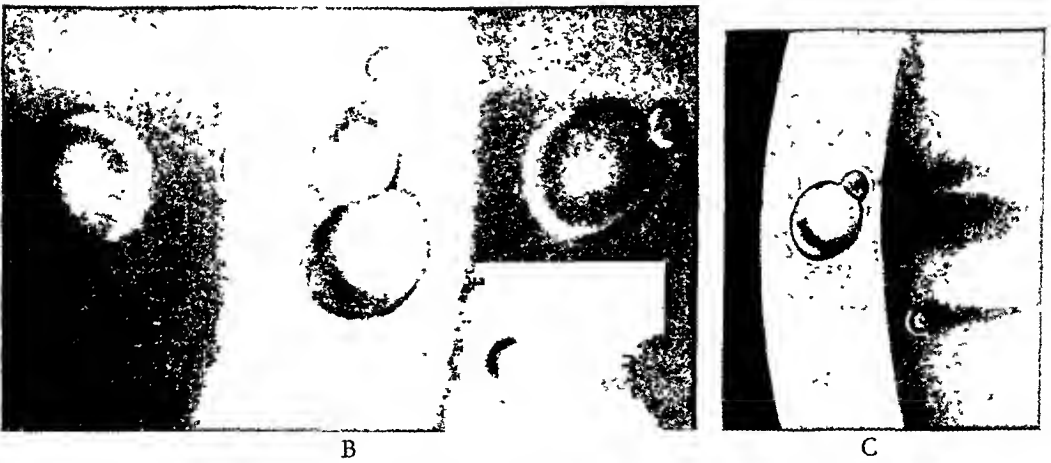
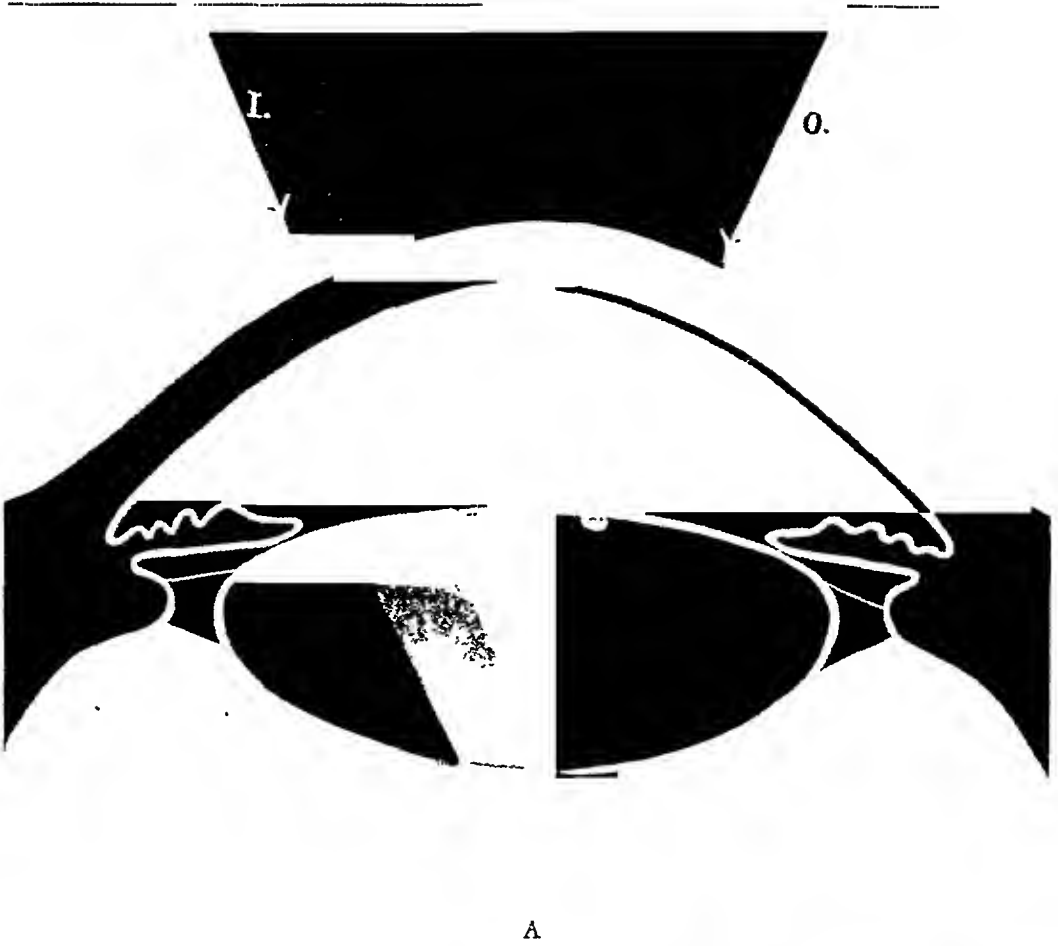


FIG. 352. A. Diagram showing technique of retro-illumination. *I*, incident beam; *O*, observation. Small subcapsular vacuoles are viewed in the light reflected from the deeper parts of the lens. B. Actual appearance of subcapsular vacuoles by retro-illumination. The two laterally-located vacuoles illustrate unreversed illumination. C. Actual appearance of vacuoles in direct focal illumination. Note shagreen area of the anterior lens capsule.

the temporal to the nasal or vice versa, one obtains sagittal "serial sections" of the lens. This is repeated superiorly and inferiorly to cover the entire exposed extent of the lens insofar as the pupillary diameter permits. In the peripheral areas it will be noticed that the zones of discontinuity are not exactly concentric but tend to diverge from each other owing to the greater curvature of the internal nuclei as compared to the curvature of the capsule.

RETRO-ILLUMINATION

By employing light reflected from the deeper portion of the lens as the focused beam passes through it obliquely, it is possible to view alterations situated more anteriorly and to one side of the beam (Fig. 352). Especially valuable as a source for retro-illumination is the light obtained by specular reflection from the posterior capsule. Although small in area, its brightness is of great advantage. Owing to the yellow color of this reflex, changes seen anterior to it will have a yellowish tinge (Fig. 353). The use of retro-illumination is very valuable for viewing anterior capsular changes, such as deposits or foreign bodies, opacities, spokes, and also vacuoles below the capsule (Fig. 352 A, B; Plate LXVII, figs. 3, 4). In opacifications of the posterior capsule and posterior cortex, e. g., posterior cupuliform cataract (saucer-shaped), the involved layers act as reflection screens. This corresponds to the use of an opaque pupil (cataract or membrane) for retro-illumination of the cornea.

Remains of the pupillary membrane (threads or stars), whether pigmented or not, are also well outlined by retro-illumination (although they are seen best by direct focal illumination). By their shape and ultimate connection to the lesser iris circle, they usually can be differentiated from pathologic deposits of pigment or exudate on the anterior lens capsule. In severe plastic iritis, vessels from the iris may cross the anterior lens capsule. Retro-illumination will bring out the red color of tiny capillaries if they contain blood. Such vessels might appear as gray lines by direct focal illumination.

The optical effects produced by this method will depend on the character of the alteration viewed; that is, whether the detail

observed in retro-illumination is (1) obstructive (opaque to light), (2) responsive (scatters light), or (3) refractile — refracts, minimizes, or distorts the views of the background.* Also the optical

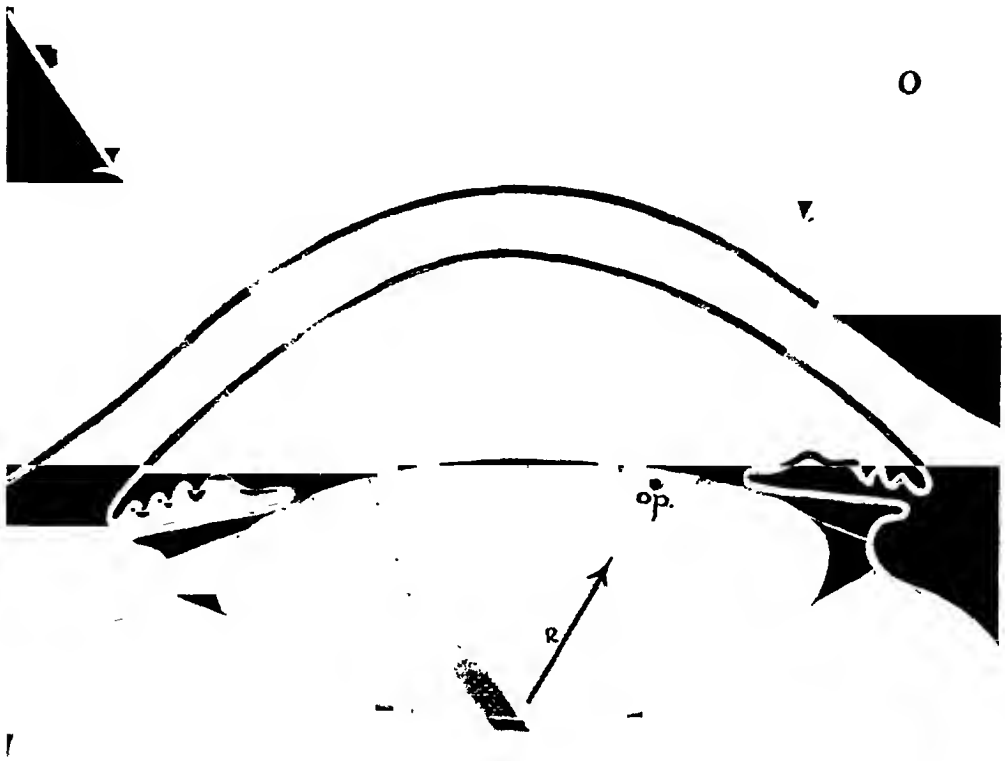


FIG. 353. Retro-illumination of the lens. Method employing the small brilliant zone of specular reflex of the posterior lens capsule as a reflecting surface. *I*, incident beam; *R*, Retro-reflected specular rays; *op*, opacity; *O*, direction of observation. (Diagrammatic.)

effects projected by certain observed details will vary depending on whether they are observed by direct retro-illumination or indirect retro-illumination. (See Vol I, Fig. 85; also Fig. 352 B.) For example, a vacuole viewed by direct retro-illumination will have a light center surrounded by a dark border. By indirect retro-illumination the vacuoles display unreversed illumination, that is, only the margin of the vacuole nearest the light is outlined by a bright crescent of light. Solid or opaque features reveal reversed illumination, that is, their side or edge away from the light will be brighter. Generally speaking, direct retro-illumination is most

* See Volume I, Chapter III, pages 83-91.

useful in observing structures that obstruct or refract light, while indirect retro-illumination is recommended for the study of structures that are refractile or respersive. In this way one may gain information not only concerning the form of structures but also about their refractive index as compared with the refractive index of the adjacent media.

SPECULAR REFLECTION

The smooth glossy surfaces of the lens capsule and nuclei act like mirrors and as such reflect incident light both regularly and irregularly (Vol. I, page 91). Similar to the cornea, when the eye of the observer is in the path of the regularly reflected rays (the angle of incident being equal to the angle of reflection) and is focused exactly on the surface, the place where this phenomenon occurs becomes visible (zone of specular reflection). The fact that we are able to discern not only the zones of discontinuity but also mirror reflecting zones on the surfaces of the inner nuclei of the lens as well as from the capsule proves, as already indicated that no uniformly gradual change in index of refraction occurs. Rather there are sudden changes, as indicated by the presence of zones of discontinuity (also called "bands") and their mirror reflexes. According to Vogt, the old opinion that the index of refraction in the lens changes during life in only one zone of discontinuity, is not borne out by factual evidence disclosed by the biomicroscope.

Mirror Reflexes of the Anterior and Posterior Capsule. As the beam is moved from side to side across the surface of the lens, a bright reflex (the shagreen) is visible (Plate LVIII, fig. 1, 2). Its glittering design is suggestive of beaten silver. Optically the design seen in the shagreen is due to the irregular reflection caused by small irregularities of the capsular surface. The detail of the shagreen of the anterior surface of the lens does not represent the actual lens epithelium and is not comparable to the posterior corneal surface where it is possible to see in the specular reflex the outlines of the endothelium. Actually, the lens epithelial cells can be seen with difficulty under high power in the central portion of the specular

reflex as much smaller dots in the design of the shagreen (Fig. 354). Vogt has suggested that this difficulty in focusing the epithelium sharply may be due to the fact that reflections from the overlying

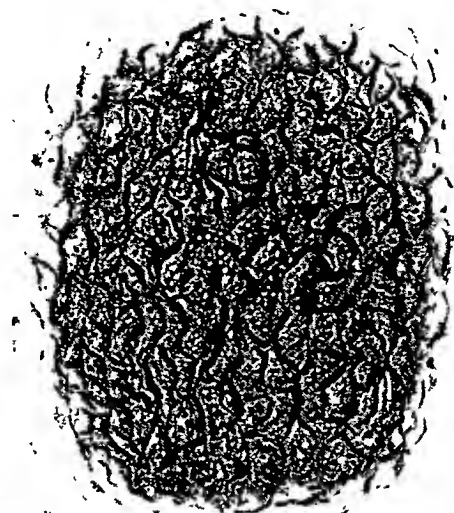


FIG. 354. Shagreen area (specular reflex) of the anterior lens capsule. Small dots probably represent reflections from the underlying epithelium (high power).

capsule cause interference. Also, the difference in index of refraction between the hyaline capsule and its underlying epithelium, as well as between epithelium and cortex, seems to be less than that between cornea and aqueous.

The epithelial cells also appear as small grooves or prominences and are best seen especially over the dark background of an underlying cortical water slit. According to Gullstrand and Vogt, the capsule, epithelium, and the superficial lens fibers with the system of lens sutures probably all contribute in the formation of the anterior shagreen. They emphasized this point because when focusing the shagreen it is possible at times to make out dark suture lines and a radial fiber design. In my opinion the reason for the appearance of the suture lines and radial fiber design in the shagreen is not that they actually go to make up the shagreen of the capsular surfaces but that when we focus the capsule shagreen, we may at the same time get internal specularity from the subcapsular layer. In other words through the capsular shagreen we see specular reflection

from the underlying surfaces. This is similar to the occurrence in the cornea of intrinsic specularity (Vol. I, page 103). Also one can easily be convinced on this point because after focusing the capsular shagreen, it always requires a minute shift in angulation between illumination and observation to bring out the suture and fiber design more clearly within it.

Consequently I agree with Butler who also considers that the anterior and posterior shagreens belong to and define the surface of the hyaline capsule. He pointed out that (1) the presence of a shagreen on the posterior capsule where the epithelium is absent indicates that it is not a necessary requirement in its formation; (2) the shagreen is present even in morgagnian cataract where the adjacent fibers are liquefied — this would tend to rule them out as a necessary component in the formation of the shagreen; and (3) in glassblowers cataract and exfoliation of the anterior layers of the hyaline capsule it is possible to observe the shagreen on the detached capsular lamellae.

As was shown in Vol. I, Chapter II, page 93, defects, such as deposits or opacities, elevations or depressions, may interfere with the specularly reflected rays and may appear as dark spots in the plane of the bright specular area.

The shagreen* area of the anterior lens capsule appears as a sharply limited zone occupying only a small part of the entire length of the surface of the opalescent lens block at any one time (Plate LVIII, fig. 3). Its width is directly proportional to the width of the slit opening. When the angle of the beam and axis of observation are so arranged that the specular reflex is not present at the point of capsular illumination, the surface of the lens block is seen by a somewhat diffused light and consequently appears gray and opalescent. If the unfocused beam (pre- or postfocal) is employed, the edges of the shagreen area are unsharp and show a gradual transition into the neighboring areas. By having the patient look slowly

* The word "shagreen" (F. *chagrin*) was adopted because of its fancied resemblance to shagreen leather, the surface of which is granular. Personally, I feel that the normal anterior zone of specular reflections of the lens more closely resemble the appearance of "beaten silver" and that the brighter and yellower reflex of the posterior lens surface simulates "beaten gold."

to the temporal side, it will be seen that the shagreen area moves to the nasal side and vice versa; if the gaze is slowly directed nasally, the temporal side of the anterior lens surface shagreen area becomes visible. The opposite maneuver holds true for the brighter, smaller, and yellower zone of specular reflection from the posterior capsule which actually is an inverted image. Because of the greater curvature of the posterior surface of the lens, the amount of specularly reflected rays from this surface at any one given point is smaller than that from the anterior surface. This accounts for the fact that the specular reflex of the posterior surface is always smaller than that from the anterior. Because the shagreen areas accurately define the capsular surface itself, they may be used as planes of reference for the purposes of localization when employing the wide beam (see traumatic cataract page 1244), e.g., in determining whether a lesion is capsular or subcapsular. Even when employing the narrowest possible beam and high power it may at times be difficult to decide whether an alteration is directly on the surface of the capsule, within its hyaline substance or even just beneath it, i.e., in the epithelium. If the alteration produces a defect (a dark or shagreen-free area) in the shagreen field, it definitely must be on/in the actual surface of the capsule.

Mirror Reflexes (Zones of Specular Reflection) of the Anterior and Posterior Adult Nuclear Surfaces. Owing to the greater curvature of the adult nuclear surfaces these reflexes are necessarily smaller. Also since these surfaces become more reflecting with age, they are seen more easily in adults than in the young. Both anterior and posterior nuclear reflexes are less bright and yellower in color than the capsular reflexes (Plate LVIII, figs. 1, 2, 3, 4). This yellow color, especially in the case of the reflex of the posterior adult nucleus, tends to become more orange or reddish with age. Since the posterior capsular reflex is always more yellow, the reddish tinge of the reflex of the posterior adult nucleus cannot be entirely caused by the physiologic increase in yellowness of the nucleus during age. In both these cases, the light filters through the yellow nucleus twice—on its way into the lens and on its passage back. The difference in

color between these two posterior specular reflexes is probably due to differences of intensity of reflection as well as to "selective" reflection (Vogt) owing to the special physical structure of the zones of discontinuity. In order to see the zone of specular reflection of the anterior surface of the adult nucleus, one first focuses the specular reflex of the anterior surface of the lens. At any given point, when this is in focus, the reflection from the surface of the adult nucleus will not be seen. In other words it is impossible to see both in the same visual axis since the curvature of the anterior adult nuclear surface is greater than that of the anterior surface of the lens. In order to see the reflex from the surface of the adult nucleus when one looks through the microscope, it will be necessary either to change the direction of the observation or to have the patient change his direction of gaze—say, slightly toward axis of illumination. Since the direction of the light beam is usually from the temporal side (we see the specular reflex of the anterior lens surface in the temporal region), the reflex from the surface of the adult nucleus comes into view as the patient turns his gaze slightly temporally. The rays specularly reflected from one given point enter only one objective of the microscope at a time. By closing one eye the observer can, with proper angulation of the beam and the microscope, see the specular reflex of the anterior surface of the lens in one ocular; then by closing the other see the specular reflex from the surface of the adult nucleus. In each case only one reflex will be seen in a single ocular.

BIOMICROSCOPIC APPEARANCES OF THE NORMAL LENS

As mentioned on page 964 the passage of the focused beam through the lens discloses well-defined relucant stripes or zones of discontinuity. These zones are best seen with the narrow beam and exact focusing of the microscope.

For the purposes of description, the zones of discontinuity of an adult lens (as seen with the optic section) from before backward (Fig. 355) may be enumerated as follows: (1) anterior capsular stripe; (2) anterior line or stripe of disjunction; the intervening

layer between this stripe and the next consecutive one (adult nucleus stripe) representing the anterior cortex; (3) anterior stripe of the adult nucleus; (4) anterior outer stripe of the fetal nucleus (outer



FIG. 355. View of the zones of discontinuity in the lens (left to right, 1-10) as seen by means of the optic section. (Diagrammatic.)

embryonal stripe of Vogt);* (5) anterior inner stripe of the fetal nucleus; the dark central interval (the truly embryonal part of the lens): (6) posterior inner stripe of the fetal nucleus; (7) posterior outer stripe of the fetal nucleus; (8) posterior stripe of the adult nucleus; the posterior cortex: (9) posterior linear stripe of disjunction and (10) posterior capsular stripe.

The biomicroscope not only permits observation of these stripes and intervening areas but also their characteristic markings, such as their zones of specular reflection which differ one from the other

* Occasionally additional stripes located between 3 and 4 and between 7 and 8 are seen. I have designated them as adolescent. See Fig. 364.

as regards size and color, their suture systems, and, on the posterior capsule, the remains of the hyaloid vessels and canal.

It will be recognized that each stripe represents optically a section through a zone of discontinuity or place where a sudden change in the index of refraction occurs. Since we assume (and with good reason) that these zones, especially the internal ones, represent areas corresponding to the nuclear surfaces, then the less relucient areas lying between these zones represent sagittal sections through the thickness of the nuclei. The dark interval, the most homogeneous and least relucient part of the lens formed in embryonal life from fibers developed from the epithelium of the posterior part of the lens vesicle (embryonic nucleus), therefore lies within the fetal nuclei. This tends to divide the lens equatorially into an anterior half and a posterior half. Clinically, the dark interval may be a place of lesser resistance as evidenced by the occasional appearance of two nuclei in extracapsular cataract extraction. In addition, in biomicroscopy (of) nuclear cataracts, the middle part of the dark central interval may remain uninvolved leading to the so-called "lens of double focus" (page 1150). The curvature of the posterior capsule, as is well known, is greater than that of the anterior capsule. Correspondingly, the curvature of the posterior stripes is greater than that of the anterior. The radii of curvature of the stripes decreases as the center of the lens is approached. Hence the curvature of the stripes increases the closer they are to the central interval. Also the stripes are not concentric to one another, owing to the fact that the distance between them is less in the axial region and gradually widens toward the equatorial regions. This results in what Vogt has termed a peripheral divergence, an arrangement which, in association with the progressive lengthening of the suture systems, causes the growing lens to become less spherical or flatter. Actually the divergence of the zones of discontinuity in the lens periphery is caused by the fact that as the zonule becomes tense, the cortical sutures become longer and the caliber of the individual radial lens fibers making up the nuclei between the zones of discontinuity, especially of the soft cortex, are thicker sagittally in the periphery than in the axial regions.

Evidently the zones of discontinuity form in consequence of the growth of the lens and biomicroscopically leave indelible signs of this growth from the earliest embryonal beginnings until death. The exact rhythm of lens growth and formation of the zones of discontinuity is still unknown. It may be that the zones of discontinuity are expressive of transient inhibitions in the rhythm of lens growth. The middle part of the lens (fetal portion) is the oldest. The peripheral portions, which grow around this central portion, develop chiefly after birth; hence the youngest portions are those subcapsular (anterior parts of the anterior cortex and the deepest parts of the posterior cortex). Even in infants where the fetal (Y) sutures are closely subcapsular, lens substance between the sutures and the line of disjunction is already to be seen. This is the earliest indication of the so-called future adult nucleus and cortex, the surfaces of which become increasingly more reflective with age (page 1019). As already mentioned the biomicroscope permits us to see at a glance the "life history" of the lens and by using the zones of discontinuity as reference points not only to localize the position of normal and pathologic changes but also to determine at what period in the "life history" of the lens they formed. For example, just before birth the lens consists of the central (fetal and embryonic) nuclei surrounded by the capsule. The greater part seen later between the fetal nucleus and the capsule has of necessity grown in after birth.* So it is easily understood that congenital opacities will have to be found within or near the central nuclei (embryonic and fetal) or connected to or in the vicinity of the lens capsule (polar cataracts) since these were the only parts of the lens extant during embryonic and fetal life. With certain exceptions (e.g., nuclear cataract, which appears to start in the fetal nucleus), changes seen developing outside the central nuclei ordinarily cannot be considered as congenital in origin, although they may be hereditary (abiotrophic) in character. An instance of this is seen in the case of coronary cataracts, undoubtedly genetically determined, which appear after

* In an infant aged two months there already was a small amount of lens substance between the capsule and the surface of the anterior fetal nucleus.

birth outside the adult nuclei. Also, as Vogt has shown, particularly in his studies of uniovular twins, certain presenile and senile lens changes leading to cataracts appear with such exactitude of form, location and time in each twin that it is certainly beyond the realm of chance or coincidence.

The exact significance of the zones of discontinuity is not known. Together with the pulling effect of the zonule, the suture systems and the zones of discontinuity are probably the phylogenetic expression of a mechanism intended to insure the "flatness" of the lens shape; to permit it efficiently to perform its function of accommodation. It well may be that the zones of discontinuity result from changes in metabolism, or from sudden spurts or retardation in growth rhythm. The avascular lens has an auto-oxidative system, receiving its nutrition via the capsule from the surrounding fluids. Perhaps during certain epochal periods as new young fibers develop, the metabolic rate in the older and deeper fibers degrades to a lower order resulting in physical and chemical variations sufficient to cause a change in their index of refraction. It is also conceivable that toxins might cause a similar change in the growing parts of the lens. Koby states: "An interesting phenomenon is the appearance of accessory bands (zones of discontinuity) usually in the deep cortex, and in certain cases we have been forcibly impressed that these bands (stripes) were brought on by endocrine disturbance."

The fact that there were reflecting layers in the lens was demonstrated years ago by the Sanson-Purkinje images (regular reflection). Since with the biomicroscope we now are able to see the details of the zones of discontinuity or the actual places where the reflex forms, we are no longer interested in mere optical imagery. Using a wide beam without the binocular microscope, Gullstrand was the first to describe the zones of discontinuity. Vogt (1917-1918), who discovered the value of the narrow beam (optic section), was able to describe these zones with greater accuracy. By means of the optic section he first showed the presence of the embryonal nuclei (embryonal and fetal, according to later terminology) and the markings (suture lines, hyaloid vessel remnants) on the other zones

of discontinuity (capsule and adult nucleus). In the latter part of the last century several workers noticed two small grayish reflecting zones in the vicinity of the Sanson-Purkinje reflexes which they cor-

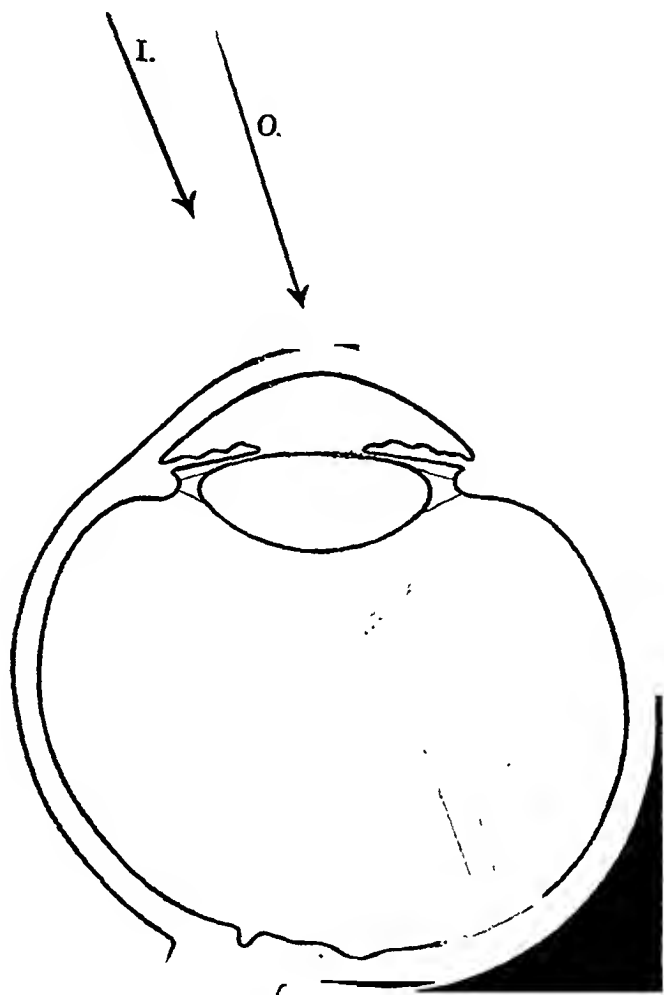


FIG. 356. Diagram showing technique of retro-illumination by means of light reflected from the fundus. *I*, incident light; *O*, angle of observation. Certain changes in the pupillary border of the iris and lens will have a red glow when viewed in this light.

rectly referred to as the surfaces of the nucleus (adult). This was corroborated later by v. Hess who showed that these were physiologic in the aged and called them the anterior and posterior miniature nuclear pictures. However, as Vogt has pointed out, the descriptions by v. Hess and even by Gullstrand of the stripe of the adult nucleus show that in reality what they saw was the posterior fetal nuclear surface.*

* Gullstrand,⁴⁶⁴ in speaking of the zones of discontinuity referred to them as the "Hess nuclear pictures." According to von Hess, the distance of the second maxima (the first

RETRO-ILLUMINATION OF THE LENS BY MEANS OF LIGHT
REFLECTED FROM THE FUNDUS

Under certain special conditions, i.e., close approximation of the axes of observation and illumination or better observation along the

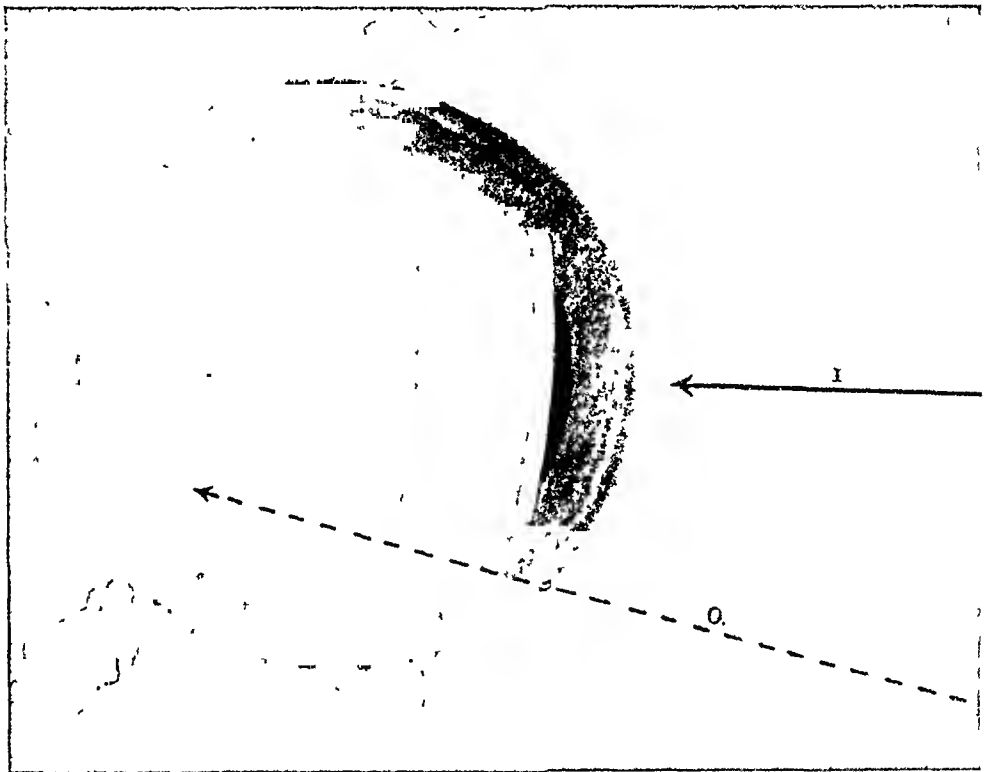


FIG. 357. Appearance of red glow from fundus when the angle of observation almost coincides with the axes of the emergent rays from the fundus.

beam axis with the unaided eye, it is possible to view certain changes in the lens by means of the red glow reflected from the fundus oculi (Figs. 356, 357). Thus, e.g., it is feasible to retro-illuminate the contents of a "spoke" in the cortex. This method corresponds to the old technique employed in ophthalmoscopic scrutiny of lenticular opacities in the light reflected from the fundus. Application of this

was the surface of the adult nucleus) from the posterior surface of the lens appears to be about two or three times the distance between that of the first maxima and the anterior capsule. In reality this would place the second maxima in the region of the surface of the posterior embryonal nucleus. Von Hess was not aware of the fetal or embryonal nuclear surfaces. As Vogt has said, Gullstrand just had to make this error because he did not know of the principal of narrowing the beam, especially since the surface of the posterior embryonal layer is the most strongly refractive layer of all the posterior zones of discontinuity.

method has been referred to already in the section on the iris and will be dealt with again (page 1317).

ANTERIOR CAPSULE STRIPE

As we observe the passage of the focused beam into the lens through a moderately dilated pupil, we see an illuminated oblique sagittal section extending through its thickness (Vol. I, Fig. 66). This illuminated portion is known as the lens block or "phantom" (Koby) and is the optical expression of the Tyndall phenomenon in the lens. The term, optic section, is reserved for the "phantom" obtained by the narrow beam (Vol. I, Fig. 70). As is the case in the cornea the width and height of the illuminated section depends on the width and height of the diaphragm opening. As the width of the slit opening is narrowed, the width of the section decreases with a corresponding lowering of its brightness. The lens area surrounding the illuminated block or optic section is dark. The contrast between the illuminated section and the adjacent dark areas is of great importance in emphasizing the visibility of microscopic details. An analogous phenomenon is seen in the dark-field type of microscopic illumination. (Also see Plate LVIII.)

The line of impact of the block or section represents the illuminated portion of the anterior capsule. With the very narrow beam this becomes converted into a line of illumination. Consequently when studying the surface of the anterior capsule the wider beam gives a larger surface area of illumination. As the beam is moved horizontally across the pupillary area, the surface of the lens block (anterior capsule stripe) appears uniformly opalescent* until the normal anterior mirror region (zone of specular reflection) is met. This occurs when the direction of the light and observation (angle of incidence equalling the angle of reflection), is so placed that the observer is in a position to perceive the rays regularly reflected.

* In older persons, frequently a delicate whitish *moiré*-like (water markings) design is seen on the surface of the anterior capsule. It takes the form of barely visible irregularly coursing lines. Whether this is related to the more marked flocculent desquamation of the zonular lamellae seen in age as well as in glaucoma capsulare is not known.

Then rather suddenly, the bright beaten-silver appearance is seen (shagreen). This mirror region occupies only a small part of the length of the anterior capsular stripe even with the wide beam, but its width extends across the full horizontal extent. Hence the lateral edges of the mirror zone are sharp, corresponding to the sharp borders of the broad capsule stripe. If the unfocused beam, pre- or postfocal (corresponding to diffuse illumination), is employed, then the lateral margins of the shagreen picture show a gradual transition into the adjoining non specular areas. Owing to the fact that we are looking at zones of specular reflection from curved surfaces, the size of the mirror area will depend on the radius of curvature of the reflecting surface. The shorter the radius (greater the curvature) the less will be the number of specularly reflected rays projected into the observer's eye at any one point. The smaller size of the shagreen area of the more curved posterior capsule is an illustration of this.* However, the smaller mirror zone of the posterior capsule is more brilliant than is that of the larger anterior one.

DETAILS OF ANTERIOR MIRROR ZONE (ANTERIOR ZONES OF DISCONTINUITY)

Shagreen. Examining the mirror area in sharp focus the coarse design (made up of grooves and prominences) is easily apparent even with lowest powers of magnification (Fig. 354). Unlike that which is seen in the zone of specular reflection of the posterior corneal surface where the outlines of the endothelial cells are perceived, in the lens this larger design is not caused by the outlines of the capsular epithelium but rather by defects in the surface of the hyaline capsule itself. Using the pre- or postfocal part of the beam or weakening the light of the wide beam and studying the anterior shagreen fields in the axial regions of the lens, it is possible to see in them portions of the sutures and a distinct radial fiber design (page

* In animals where radii of curvature of the lens surfaces are greater (spheroid type of lens) the zones of specular reflection are smaller than in the flatter lens of man. Following an iridectomy in a 58-year-old woman who was using pilocarpine- eserine mixture, Vogt found that the shagreen field became as small as that seen in a rabbit (spheroid lens). Evidently a relaxation of the zonule and with it an increase in the curvature of the lens surface occurred, owing to ciliary muscle spasm.

972 and Fig. 358).* The reflex from these fibers ends at the suture, which in itself appears as a dark stripe. Vogt⁶⁵⁰ demonstrated this point in 1921 and compared the apparent crestlike appearance of

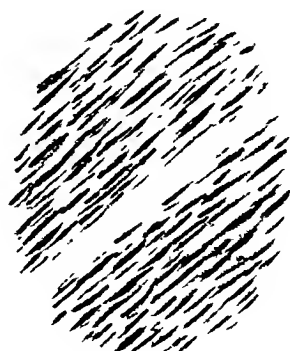


FIG. 358. Underlying suture and fiber designs as seen in the mirror zone (specular reflexes of the anterior and posterior capsule).

the sutures to that of the ribs of an opened umbrella. Although the fiber arrangements on both sides of a suture are anatomically symmetrical, the appearance is different in specular reflex because with the illumination directed from one side the reflections from these symmetrical abutting fibers are necessarily dissimilar. Since the specular reflex of the fibers is derived mainly from their surfaces and not the fiber extremities (at the suture), the reflex at border regions of the sutures is light deficient and causes the suture itself to appear as a dark line; according to Vogt's description as dark crests. The latter also noted what appears to be a slight depression or groove in the shagreen field of the anterior capsule, concentric with and just at the margin of the pupillary edge—where the posterior surface of the iris normally contacts the capsular surface. This condition can only be seen in the young where it forms a slightly depressed concave band parallel to the pupil. The fact that the contact of the iris could produce such an impression speaks for the softness of the capsule and the underlying fibers and also might explain the reason for the location of Vossius' traumatic ring opacity

* In my opinion these features are not directly related to the capsular shagreen but result from intrinsic specularity of the fibers below it. As in the case in the cornea, when viewing the anterior mirror zone one may at the same time see specular reflexes behind it (Volume I, page 103).

seen in the young. In addition, the shagreen fields, as would be expected from the fact that the thickness of the capsule varies from place to place between the pole and equator, differ as one observes it in different areas. In some sectors the fiber design may not be sharply visible. As a matter of fact in the polar region the fiber design is frequently seen better when the posterior or prefocal part of the beam is employed, because as previously mentioned their reflection is independent of that of the shagreen of the hyaline capsule. However, it should be remembered that what is seen in the zones of specular reflection depends on the angle between illumination and observation, and the appearance of details will vary as we alter this ever so slightly. In addition, generally speaking just as the zones of discontinuity are better differentiated in axial regions as compared to the far periphery so are all their mirror zones. As a matter of fact the reflexes of the latter are not seen peripherally.

Epithelium. Using a higher magnification, $35\times$ to $60\times$, the outlines of the epithelium cells themselves may be seen as smaller irregular glints within the design of the coarser shagreen and only within limited areas (Fig. 354). The difficulty in seeing the outline of the lens epithelium clearly in the zone of specular reflection as compared to the corneal endothelium is probably due to the fact that the light reflected from the more reflecting capsule in front blurs them somewhat. In addition (Vogt) the difference in index of refraction between the epithelium and the capsule as well as between the epithelium and underlying cortex seems to be less than that between the cornea and the aqueous. The margins of the epithelium cells are best seen in the young, especially with the aid of the cylindrical light bundle. They do not appear as a well-formed mosaic as is the case with corneal endothelium, but for the fact mentioned are seen as irregularly outlined reflecting dots, a few in each shagreen field.

Shagreen Spheres. This term, originated by Vogt, applies to small spheric to ovoid bodies from 20 to 60 μ in diameter seen in the anterior mirror zone of the lens toward the periphery (Plate LIX, figs. 5, 6). Their exact significance is unknown. In order to be seen maximum dilation of the pupil is necessary since they are usually

PLATE LIX

FIG. 1. Suture lines (slightly dilated) seen at the level of the adult nucleus in an old person. Diffuse illumination.

FIG. 2. Suture lines (same case as Figure 1) as seen by retro-illumination, by light reflected from the deeper parts of the lens.

FIG. 3. Suture system seen by diffuse illumination in a man of 50 years.

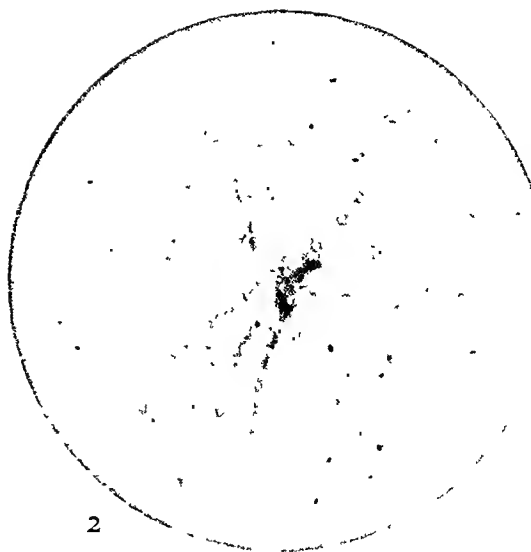
FIG. 4. Same as Figure 3 viewed by direct focal illumination.

FIG. 5. Relief and suture system of the surface of the adult nucleus. Showing shagreen spheres in the specular reflex. Diffuse illumination.

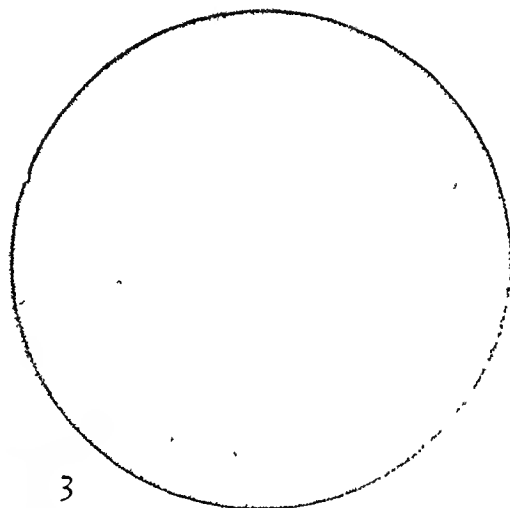
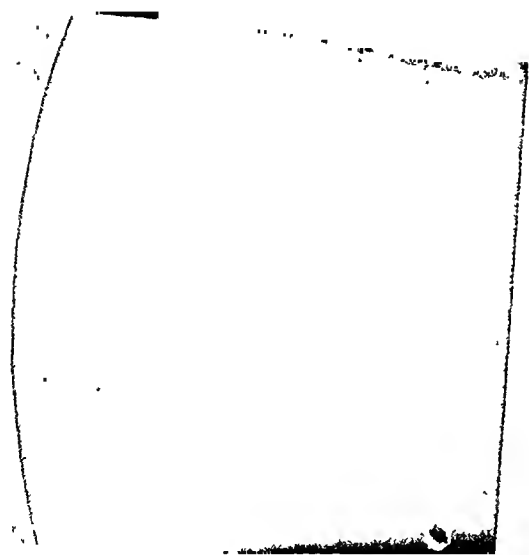
FIG. 6. Same as Figure 5 as seen by direct focal illumination.



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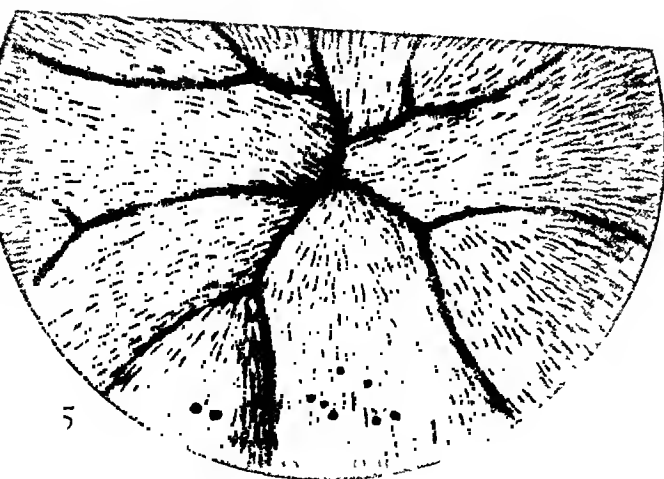


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4



5



6

found in a zone corresponding to the middle and peripheral one-third of the visible anterior lens surface; occasionally they may be found more axially. The wide beam and lowered illumination should be employed in order to veil the brilliancy of the reflection of the capsular moiety of the shagreen. The spheres appear as black holes or spots in the shagreen field. By indirect illumination (at the side of the mirror zone) a glistening mirror reflex from them proves that they are not artifacts and also shows their spheroidal shape. They are rarely seen in the young, but in adults and the aged they are constant and presumably physiologic findings. They are best seen in older persons with cataractous lenses. Unlike vacuoles, shagreen spheres cannot be seen with the ophthalmoscope nor by means of retro-illumination using the reflected light from the mirror zone of the posterior capsule. They may represent verrucosities of the lens fibers.

Faint Iridescence (Color Display) of the Anterior Mirror (Shagreen) Zone. When observing the anterior lens mirror region of adults or of the aged, one is frequently struck by the presence of a rather faint color display not unlike that seen when oil is floated on water (Plate LVIII, figs. 1, 3). The tints vary from pale green to a reddish yellow. This type of iridescence is also found in the posterior capsular shagreen. This should not be confused with the more marked iridescence seen in complicated cataract (pages 1169, 1285) or in cases of chalcosis lentis wherein copper is deposited in the subcapsular layers following the intra-ocular introduction of a cuprous foreign body. In his earlier work Vogt stated: "This physiologic iridescence may be due to delicate remnants of the embryonal capsular membrane," while later he stated that it may be due to the interference phenomena caused by a very thin layer of fluid less than 0.2μ , basing this newer opinion on his findings in cases of traumatic cataract. Evidently this display of color must optically be due to some interference phenomenon within the capsule itself.* Very little is known

* In order to dispel any idea that iridescence was brought about by chromatic aberration or diffraction within the illuminating system, Vogt directed the beam on a white disk of porcelain. In this case color aberration occurs only at the margins of the illuminated area whereas in the shagreen or in the case of complicated cataract it occurs within the center of the illuminated areas.

concerning the morphology and physiology of the lens capsule, and it has only been since the advent of biomicroscopy that our attention has turned to it.

ANTERIOR STRIPE OR LINE OF DISJUNCTION *

This thin stripe or zone of discontinuity, which is found just behind the capsule stripe (but is also present in front of the posterior lens capsule), can be clearly identified only with the narrow beam (Fig. 355). When the width of the slit opening is reduced to 0.5 mm., the width of the beam will be 20 μ . In the axial regions or poles, the stripes of disjunction seem almost to fuse with the capsule, but with the narrowest beam an area of separation between them can always be demonstrated. They are present in the young individuals and even in infants when it is possible to examine them biomicroscopically. Peripherally the space between these stripes and the capsule gradually widens, producing the so-called peripheral divergence. Except for its peripheral divergence this line might be compared to the double line (seen only in optic section) on the anterior corneal surface. However, in the case of the cornea the second line represents the surface of Bowman's membrane. (See Vol. I, Fig. 69.) Peripheral divergence is also a feature of the deeper zones of discontinuity. Vogt has designated this stripe as "abspaltungstreifen," the exact translation of which signified a progressive widening of separation toward the periphery. In other words the radii of curvature of these stripes diminishes peripherally. This anterior stripe is in contrast to the other zones of discontinuity less distinct with age, especially as the relucency of the cortex becomes greater. Vogt was unable to find them in cases of cataracts occurring in myotonia, tetany, and in the separation of the zonular lamellae in glass blower's cataract.

ANTERIOR STRIPE OF THE ADULT NUCLEUS

This grayish white stripe or band represents an illuminated section of the adult nuclear surface (Fig. 355; Plate LVIII, fig. 3). Since

* This zone of discontinuity was first described by Gullstrand.⁴⁶⁴ Vogt described the other zones of discontinuity in 1917-1918.

accessory faintly visible stripes may occasionally be seen in the cortex, the term "anterior stripe of the adult nucleus" refers to the deepest and most luminous one that lies between the anterior line of disjunction and the surface of the anterior fetal nucleus. Even as we narrow the beam, this illuminated area is never as sharply defined as the capsular stripe or disjunction lines; consequently except for the purposes of localization it is preferable to observe it with the wide beam or at times in older persons when it becomes more reluctant and highly reflective to study even a greater part of its relief by diffuse illumination. It should be repeated that even though some of the zones of discontinuity appear as lines or possibly surface planes in optic section, actually they have a very definite sagittal thickness. In children, where all the zones of discontinuity are less reflective, a faint stripe is found deeper to the line of disjunction anteriorly, and in front of it posteriorly (Fig. 356). Although they are called the "adult nuclear stripes" it is still not known whether they actually are the same stripes (so termed in adults). Probably they develop into them * (page 992). Already during adolescence these zones of discontinuity, when viewed with the narrow beam, no longer appear as sharp lines as for instance the capsule stripe or disjunction lines, but rather as a hazy band having a definite sagittal thickness. With age the width of this band increases, especially as it approaches the periphery (equator). So that it is always thicker here than in the axial region of the lens. The fact that this band becomes thicker and more reluctant with age (while the other internal zones of discontinuity become less reflective) suggests that it may be formed by the coalescence of several newly formed zones of discontinuity. At the end of the first decade, it is already possible to see a stripe which probably corresponds to the future highly reflecting band of the adult nucleus. Auxiliary stripes in the cortex were already noted by Vogt and Koby. Goldmann⁴⁵³ went a step farther and

* With this in mind, Vogt studied a lens of a freshly removed gliomatous eye of a 13-month-old infant. He found that the lens was very thick sagittally and that the stripes of disjunction were equidistant (no peripheral divergence) from the capsule. The zone of discontinuity, which corresponds to that of the adult nuclear stripe, was present but was much more curved. Because of its extreme curvature, he suggested that this stripe might correspond to the outer embryonal zone (fetal). In a very cooperative 2-year-old child I found this stripe outside the fetal one.

showed that with arc-lamp illumination it is possible to see a series of these lines in the cortex which increase in number with age. These lines are formed by the progressive growth of new fibers which are being continually displaced inward. With periods of retardation in growth rhythm a group of fibers may become more reflective or develop a difference in refractive index and thus optically produce a line.

This idea occurred to Goldmann after seeing faint lines (zones of discontinuity) appear in animal lenses at the place where damaged layers of fibers were receding to deeper levels and that these lines optically were the expression of transient inhibition of growth. In order to explain the genesis of the peculiar appearance of the adult nuclear bands in man he observed 114 persons of various ages. In these examinations he used the biomicroscope with an arc lamp source and a special measuring ocular. In 66 persons with normal lenses he made the following observations: (1) The distance between the disjunction line and the capsule does not show any definite relationship to age. (2) The distance between the anterior margin of the anterior senile nuclear stripe and the anterior capsule decreases slightly with age. (3) The distance between the posterior margin of the anterior senile nuclear stripe and the anterior capsule increases markedly. (4) The most striking change is the increase in thickness of the so-called "band" of the anterior adult nucleus.

This would indicate that the apparent increase of the reflective power of the adult nuclear band with age is due to its great increase in thickness. Examination with the arc source of illumination and narrow slit opening revealed that the adult nuclear band was composed of a series of fine lines. Similar lines were also found in the cortex and extended up to the disjunction line. In all cases these auxiliary lines were seen more readily at the periphery than axially. The number of the lines or zones of discontinuity increased with age. In a person 20 years of age, he found only three or four stripes while in an individual of 80 years he found as many as twenty. However, these auxiliary or elementary lines were not visible in every case even with the arc source; in most instances, it is not possible to

see them with the ordinary nitra bulb light source. Goldmann estimated that approximately one elementary line is produced every two or three years. According to this, elementary lines are laid down

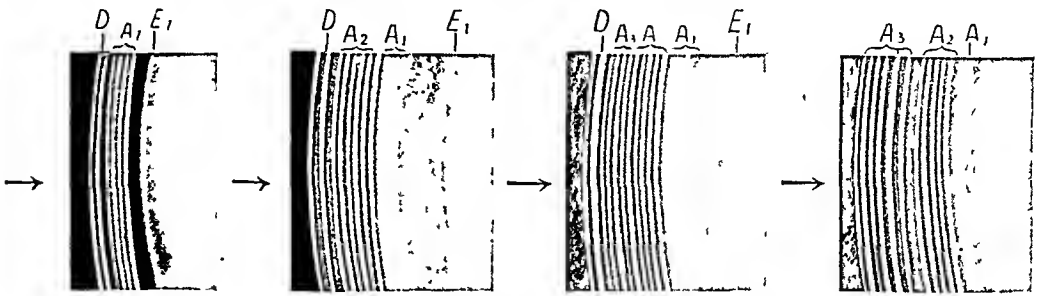


FIG. 359. Diagram showing accessory stripes in the formation of the band of the adult nucleus. (After Goldmann.)

subcapsularly and with age the older ones are pushed deeper.* Owing to the increase in number and to coalescence, the sagittally thickened and increasingly reflective band of the adult nucleus results. In other words, the increase in reflection of the adult nuclear surface results from the increase in number of elementary lines, thus producing a reflecting layer of greater thickness, rather than an increase in reflecting power from single layers. Apparently these lines are not the expression of a special kind of fiber which has a rhythmic growth but rather a phase through which every fiber has to pass. There is also the possibility that the mechanism of internal lenticular accommodation plays a role in the formation of these lines. With age, the anterior border of the thickened adult nuclear stripe progressively approaches the capsular stripe. Hence the distance that the newly formed lines have to travel is progressively less. This process in which the nucleus increases in thickness at the expense of the younger new fibers of the cortex, may progressively limit the latter in performing the act of accommodation.

According to Goldmann's investigations it is possible to differentiate three zones of elementary stripes which correspond to the

* Goldmann suggests that the following experiment will give one an impression of this mechanism: "If one puts many cellophane plates, one on top of the other, fills the interspaces with alcohol and squeezes them between two microscope slides, and then observes them with the narrow beam of the slit lamp, one will have a picture of a large zone of reflexes like the senile nucleus band in which single elementary lines are visible, but nearly always fewer than the number of plates. The same plates observed in diffuse illumination show the dull luster which is very similar to the inner reflection of old lenses."

different periods of life (Fig. 359). The first group which he designates as A_1 are seen early in life (before the twentieth year) and are located just behind the disjunction lines. A dark interval separates

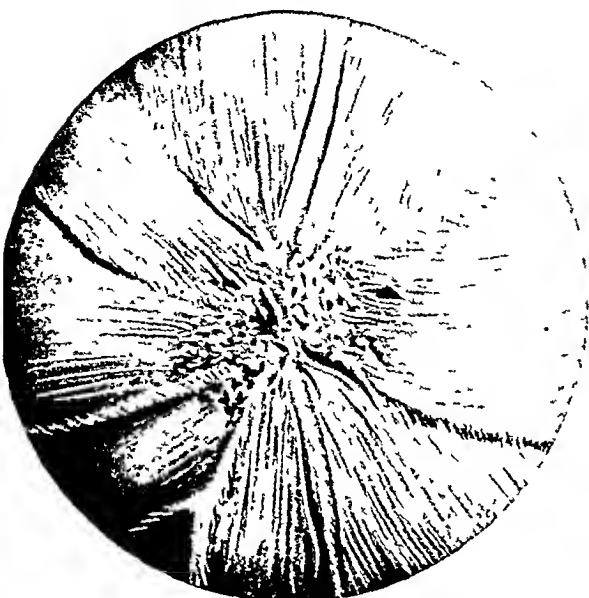


FIG. 360

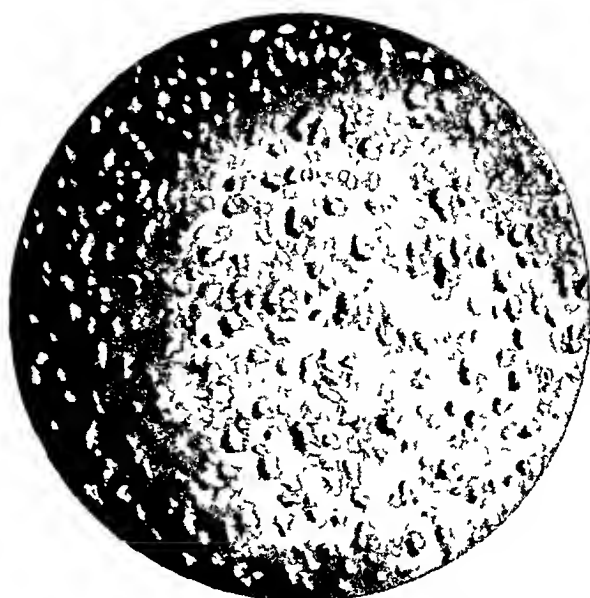


FIG. 361

FIG. 360. Ridges of the adult nuclear relief. (After Vogt, from plastic model.)

FIG. 361. Knobs of the adult nuclear relief. (After Vogt.)

them from the outer embryonal (fetal) nuclear zone. Between the twentieth and fortieth years another series of lines appears in front (capsule-ward) of A_1 . He calls this group A_2 . By this time the deeper group A_1 becomes fainter, grayish and dull, so that its component lines are no longer easily seen. It may be that these join with the outer embryonal nucleus. Toward the end of the third decade the number of lines in A_2 increases (the zone getting thicker) and become increasingly brighter and yellowish. It is the condensation of this group (A_2) which forms the highly reflecting band of the adult nucleus seen in older persons and which presents the typical relief of the adult nuclear surface (page 995). After 40 years of age, another group of lines (A_3) forms subcapsularly in the cortex. In the direction of A_2 they are not sharply demarcated, but anteriorly they are well individualized and are whitish.

With age, the anterior band of the adult nucleus becomes so relucant that it reflects more light than the capsule stripe. Also this zone develops a peculiar maplike relief, erroneously interpreted

by von Hess as the beginning of a senile cataract. This effect can easily be seen if one directs the unfocused beam (diffuse illumination) with a large angle of incidence into the wide pupil. In the axial region, the many branched suture system (from 6 to 10 main branches and from 10 to 15 sub-branches) stands out as if elevated from the surface with a characteristic design of the lens fibers (Fig. 360; Plate LIX, figs. 3, 4). In the periphery peculiarly shaped knob-like prominences are seen (Fig. 361). These round or oval elevations are never confluent. At times they reflect a little light and appear glossy. However, it should be pointed out that the relief of the adult nucleus varies in appearance from case to case. In some it is very marked while in others it is faint and blurred. The vertical ridge seen only in the axial parts is probably the primary one and hence tends to be broader and higher than its branches. Employing a wide angle of incidence of illumination and changing the direction of the light in the pupil from the temporal to the nasal side and vice versa, shadows formed on the less illuminated sides of the ridges will accent their relief and cause them to stand out sharply above the surface (Fig. 360). The ridges may be absent; in this case the sutures will appear as dark lines on a smooth surface. Frequently, when the ridges of the sutures are not seen, the surface will be marked by the presence of the knobs (Fig. 361). The design of the relief corresponds to the direction of the sutures and consequently extends itself in a continuous way to the periphery where the ridges gradually flatten out. With higher powers, reflections from the fiber design are commonly seen in the form of fine lines. These lines (bundles of fibers) run at right angles to the ridges and as they cross them become convex in a frontal direction. In other cases the center of the ridges may be depressed, forming a sort of trench. It may well be as Vogt suggested that these various appearances of the nuclear relief hint towards shrinkage of the adult nucleus. In order to show the incidence of the relief Vogt and Lüssi⁶⁶⁵ studied a series of 628 subjects. Of this group, 75 had coronary opacities, 75 had other forms of opacities, 20 had nuclear cataract and in 83 the opacities were negligible. Table XVI was taken from Lüssi.

From these figures it will be seen that the relief can rarely be distinguished before the twenty-fifth year. It should be pointed out that in very rare cases a faint relief of the outer fetal nucleus may be present; but as this does not have the characteristics of the adult nuclear relief and is found in younger individuals, there is little likelihood of confusion. The relief of the surfaces of the inner fetal nucleus is plainly marked by the erect and inverted Y-sutures.

TABLE XVI
FREQUENCY OF THE RELIEF *

AGE OF SUBJECT	NUMBER OF SUBJECTS	PRONOUNCED RELIEF	SLIGHT RELIEF
1 to 10 years	7
10 to 20 years	84	1
20 to 30 years	116	4	9
30 to 40 years	83	3	12
40 to 50 years	87	26	19
50 to 60 years	87	32	27
60 to 70 years	92	24	20
70 to 80 years	67	10	5
80 to 90 years	5	1
Total	628	101	92

* From Lüssi. 529

It should be remembered that these appearances are not opacities and, even when prominent, do not disturb vision. They cannot be seen by transillumination with the ophthalmoscope. Pathologic features such as vacuoles just below the nuclear surface tend to be larger in size and are more circumscribed than similar subcapsular changes (Fig. 352 B, C). They can always be seen as vacuoles by retroillumination (using the shagreen reflex area of the posterior capsule as a light source) and when marked may influence vision. It has been suggested that the hemispherical projections are produced by a collection of fluid.



FIG. 362. Increase in thickness of the adult nucleus as the periphery is approached.

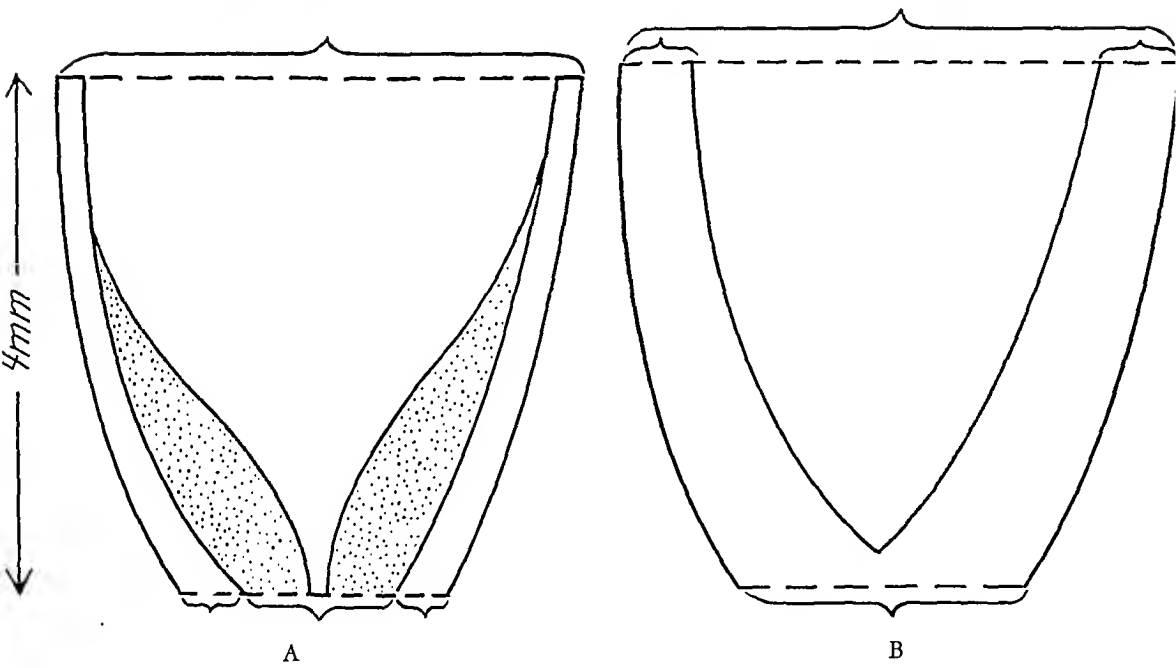


FIG. 363. Peripheral divergence of the outer zones of discontinuity. A. 21-year-old person. The more or less sharp surface of the adult nucleus in a young person widens out in a zone of increased diffuse reflection as the equator is approached. This is seen best when the narrow beam of high light intensity is projected through the lens very obliquely. As a result the nucleus equator becomes drawn out similar to the neck of a bottle. The widening of this zone (peripheral divergence) is caused by the traction of the zonule. This appearance is not found in cases where zonular traction is absent, e.g., sphereophakia. B. 77-year-old person. The surfaces of the adult nucleus do not widen and become diffuse as they approach the equator. Hence the condition seen in A is absent. Diagrammatic. (After Vogt.)

The surface of the adult nuclear stripe (especially in the young) blends into a zone of increased diffuse reflection as it approaches the equator. Here it thickens sagittally so that often it cannot be recognized as a surface (Fig. 362). In the aged this zone of increased diffuse reflection does not occur (Fig. 363 B). In the axial region the sagittal thickness of the adult nuclear stripe becomes thinner. This is of optical significance in the pupillary portions of the lens and of lesser optical importance in the periphery where, due to widening, there is a more gradual transition of refractive index. According to Vogt this increase in thickness peripherally has to do with the pull of the zonule since it is not seen in spherophakia, a condition characterized by the absence of zonular tension. Another probable instance of the effect of the zonular pull is morphologically evidenced in the normal lens by the augmented tapering of the adult nucleus in the equatorial zone. Particularly in the young both anterior and posterior surfaces approach each other to taper out and almost join, being separated or split as it were by an extension of the central interval (Fig. 363). The peripheral divergence of the adult nuclear stripes should be noted. This, together with the shorter radii of curvature of the nucleus, causes a greater sagittal thickness of the cortex in the periphery as compared to the axial region. This thickening in the periphery (manifested by the peripheral divergence of the zones of discontinuity) results in a flattening of the lens surface. In this way the shape of the lens is more disklike and only with relaxation of the zonule can its sagittal diameter increase (process of accommodation).

Mirror Zones (Specular Reflectional) of the Adult Nuclear Surfaces. Both surfaces, anterior and posterior, of the adult nucleus exhibit mirror reflexes or zones of specular reflection. Because of the smaller radius and consequent greater surface curvature, the mirror zones of the adult nucleus are smaller than those of the capsule (anterior). Not only are they smaller in size but are also less bright in intensity. The anterior mirror zone of the adult nuclear surface is yellowish white in color, and its posterior one, especially in the aged, is yellow-orange or even reddish (Plate LVIII, figs. 2, 4). Owing to the difference in curvature between the capsule and surfaces of the adult nucleus it is impossible to see the reflexes, simultaneously

with the same angle of observation. One first focuses the anterior shagreen and by changing only the angle of observation (the angle of illumination is not changed) ever so slightly in the direction of the shagreen the mirror zone of the adult nucleus will be seen. As this comes into view the shagreen is no longer seen. However, owing to the fact that the specularly reflected rays from any one given point can only be perceived in one ocular, alternate opening and closing of each eye will frequently bring them into view successively without changing the actual angle of the microscope. The same will hold true in a reverse way when viewing the posterior shagreen and the mirror zones of the posterior adult nucleus surface. As Vogt has pointed out, the reddish color of the posterior adult nuclear layer is not entirely caused by the yellow color of the posterior portions of the lens (physiologic with aging) because then the color of the posterior shagreen should also be of the same reddish hue which is not the case; instead this is a bright yellow. The light of the posterior capsular shagreen passes through the lens twice. Evidently the zones of discontinuity have special differences of intensity of reflection and are "selective." In the normal eye, no special design or features can be made out in the zones of specular reflection of the surfaces of the adult nuclei.

ANTERIOR OUTER STRIPE OF THE FETAL NUCLEUS (OUTER EMBRYONAL STRIPE OF VOGT)

Surrounding the opposed coffee bean-shaped inner fetal nuclei the surfaces of which are so characteristically marked by the Y-sutures, are the outer stripes of the fetal nucleus (Figs. 351, 364; Plate LVIII, fig. 6). In adults and in the aged, these stripes become increasingly difficult to recognize as definite zones of discontinuity. As previously mentioned, their relationship to the so-called adult nuclear stripes as seen in the young and to the inner fetal nucleus is not precisely known. Since the time of formation or development of the anterior and posterior outer stripes of the fetal nucleus has not been established it is not entirely correct to call them "infantile" stripes, as has been done by some writers. All we can say at present is that they form either just before or just after birth. From all the available

material at hand and from my own observations, I believe that they are present at birth and consequently I prefer the term "outer fetal stripes." Actually, birth creates no sudden change in the develop-

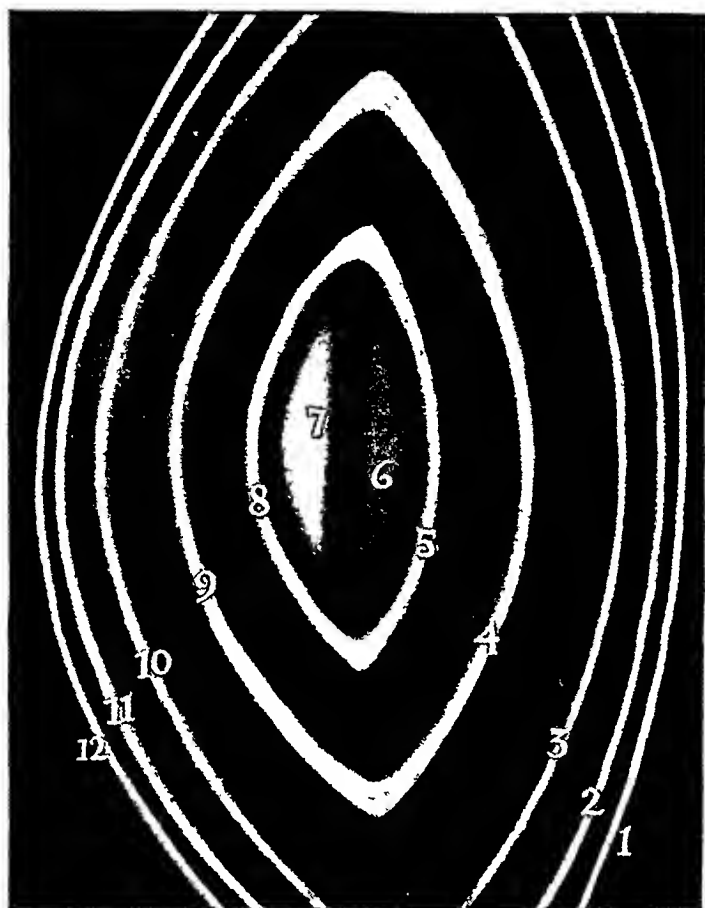


FIG. 364. Diagrammatic representation of the zones of discontinuity as seen in optic section (1-12). 1, Anterior capsule stripe; 2, anterior disjunction stripe; 3, anterior adult nucleus stripe; 4, anterior adolescent stripe; 5, anterior outer stripe of the fetal nucleus; 6, anterior inner stripe of the fetal nucleus; 7, posterior inner stripe of the fetal nucleus; 8, posterior outer stripe of the fetal nucleus; 9, posterior adolescent stripe; 10, posterior adult nucleus stripe; 11, posterior disjunction stripe; 12, posterior capsule stripe.

ment of the lens. The attempt to classify the zones of discontinuity located outside the inner fetal nuclei, as infantile, adolescent, adult or senile is not justifiable because it gives the impression that these zones form suddenly during the different epochs of life. Actually all that happens is that with age and growth of new fibers some are accentuated, e.g., adult nuclear stripes, and the others either fuse or because of increased relucency of the intervening layer become less obvious as zones or places where a sudden change in the index of

refraction occurs. In the newborn, the fetal nuclei are separated from the capsule only by a comparatively thin area, occupying relatively a much greater portion of the whole lens than it does in adults. In spite of the fact that with the growth of new fibers between the fetal nuclei and the capsule and the resulting increase in thickness of these layers (adult nucleus and cortex), the total sagittal thickness of the whole adult lens is hardly greater than that of the newborn. This would indicate, according to Vogt, "that the embryonal nucleus (fetal) suffers an important decrease in size by shrinkage during life." Koby states that these stripes (anterior and posterior outer fetal nuclear) "although narrow, are nevertheless sharp, and central to the sutures there is an increase in optical density, not sufficiently sharp and localized to be considered as a band of discontinuity, so that it is rather a question of a very diffuse zone. With age the peripheral (outer) and central (inner) bands gradually fuse." Whether this assumption is correct will only be proven when the growth of the lens in the young is more systematically studied with the biomicroscope. The other possibilities are that these structures either merge or become the "adolescent" stripes or that as the relucency of the entire adult nucleus increases the change in index or refraction at these points no longer is sufficient to produce a zone of discontinuity. Rarely in the young faint zones of discontinuity between the outer stripes of the fetal nucleus and the adult nuclear stripes may be found. These so-called adolescent stripes are usually not seen in adults and even in the young are inconstant (Figs. 351, 364). When present both these stripes show in the equatorial regions the phenomenon of peripheral divergence. The outer stripes of the fetal nucleus are more often present than this rare supernumerary line.

ANTERIOR INNER STRIPE OF THE FETAL NUCLEUS

(INNER EMBRYONAL STRIPE OF VOGT)

In the central part of the lens are the two coffee bean- or biscuit-shaped nuclei, indicated biomicroscopically by the anterior and posterior inner fetal stripes.* Between them is the so-called "dark

* As mentioned on page 976 the fetal nucleus, the core of which is the dark interval or embryonic nucleus, consists of that part of the lens that is bounded by fetal Y sutures. Koby states: "These points of references are much more precise for the fetal nucleus than the

interval," an area whose relucency almost corresponds to that of the normal aqueous. The coffee-bean like appearance of these stripes never varies and represents optically a sagittal section through the

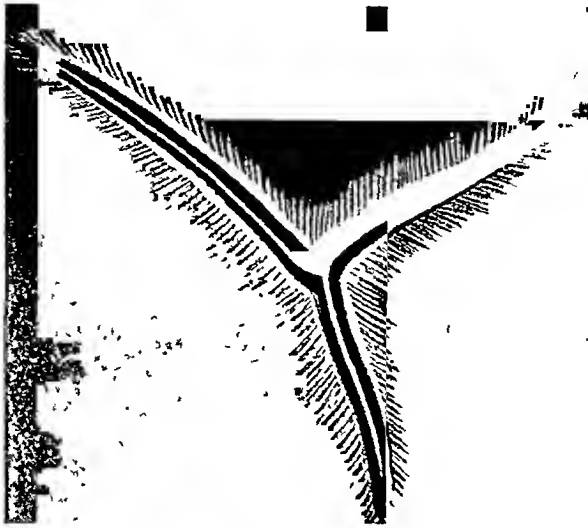


FIG. 365. Anterior fetal Y suture.

thickness of the inner fetal nucleus. The location of the nuclei is marked by the presence of the well-known Y-sutures. The anterior inner stripe of the fetal nucleus, with its erect Y-sutures, is much less relucant than the posterior one and consequently is somewhat more difficult to see. After obtaining the posterior stripe, easily localized by the inverted Y-suture, one focuses in the slightly relucant area in front of it (zone of anterior inner fetal nucleus), and with slow gentle oscillations of the beam, suddenly the fainter anterior upright Y will be discerned. It stands out, in youth, whitish against the less relucant dark background. (Figs. 345, 346.) This whitishness is the result of reflections from the feathery fibrillary appearance of the extremities of the lens fibers. As the internal relucency of the lens increases with age these sutures appear dark

bands (stripes of discontinuity)." The fetal nucleus consists of that part of the lens which embryologically develops after the completion stage of the primary vesicle. Its fibers develop from the capsular epithelium at the equator and grow around those which in the primary lens vesicle extended forward from the original epithelium of the posterior capsule, to fill its cavity. The secondary or fetal fibers in their development grow in and separate the capsule from these original sagittally directed ones of the primary lens vesicle. As time goes on the outlines of the primary lens fibers become indistinct and eventually even at birth this area appears as a dark nonrelucant zone separating the two coffee-beanlike fetal nuclei. The dark interval never shows any trace of a suture system nor do the branches of the fetal sutures ever enter it.

against the brighter background. However, in nuclear cataract the fetal sutures are delineated as white lines contrasted to the surrounding uniform milky gray of the central lens areas. Generally the angle between the two main branches of the anterior Y is wider than that of the posterior, giving the anterior Y a somewhat extended appearance (Fig. 365). Also the tendency to subbranching of any of its three arms is less than that of the posterior Λ . As a rule, save for an occasional dichotomous branching of the lower limb, the arms of the anterior fetal suture do not ordinarily branch. Often the fetal sutures are inclined from the vertical and appear as if they were reclining. Also at times the relations of the two Y's are such that they stand obliquely to each other in such a way that their vertically directed arms (anterior inferior and posterior superior) are parallel to each other. The vertical arm of the anterior Y and the horizontal arm of the posterior Λ are the first to form and also are the first to show subdivisions. This corresponds to what is seen in lower forms of life, e.g., certain fish where the anterior suture of the lens is a vertical line (no branchings) and the posterior one is represented by a horizontal line only.* It is interesting to note that in this region axially (anterior inner fetal zone) Vogt found in 25 per cent of otherwise normal lenses small groups of whitish dots which cannot be seen ophthalmoscopically and which do not affect vision. (He has termed these "the anterior axial embryonal cataract" [page 1055]).

THE POSTERIOR INNER FETAL (EMBRYONAL-VOGT) STRIPE

The dark interval separates the fetal nucleus into two halves. It will be seen that the dark interval divides the entire lens in the equatorial plane into two parts which are almost identical except for variations in curvature and in certain details of markings. Similar to the two separated halves of a coffee bean, the configura-

* In his photomicrographs of rabbits' lenses impregnated with silver nitrate Vogt shows a similar arrangement. The anterior suture appears like a vertical line and the posterior as a horizontal one. In each case the fibers run at right angles to the direction of the suture line and widen as they approach the suture itself. In the development of the suture in higher animals and also in man the phylogenetically older vertical or horizontal part of the suture starts to deflect or bend. This bending is always a forerunner of branching. In other words, the suture changes its direction slightly before it branches.

tion of the surfaces of the inner fetal nucleus is convex externally and to a lesser degree somewhat concave internally.*

The posterior Λ suture, which permits biomicroscopic localization

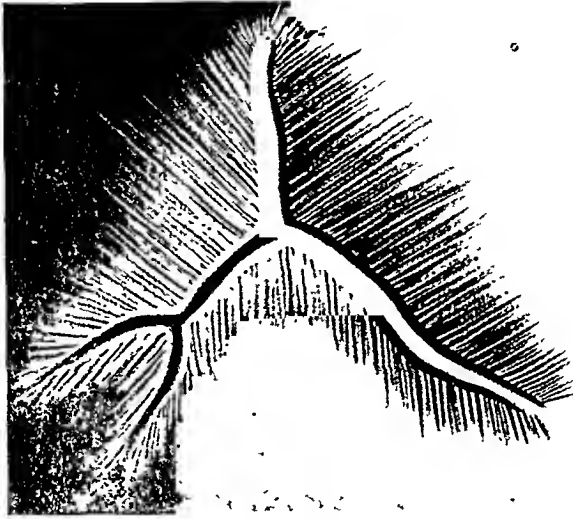


FIG. 366. Posterior fetal suture.

of the posterior inner fetal nucleus, is more easily seen than the anterior Y. As a matter of fact, in youth the posterior inner fetal stripe or zone is usually the most relucient of all, and it is wider axially than peripherally as compared to the other zones of discontinuity where the opposite holds true.

The fibrillations which represent reflections from the fiber ends as they meet to form the suture are best seen at the borders of the posterior Λ (Fig. 366). The two lower legs of the Λ join the upper vertical at an obtuse angle in the form of a gothic arch, the lower legs often extending out almost horizontally. The upper (phylogenetically younger) vertical arm only very rarely shows any branch-

* The greater curvature of the outer fetal nuclei causes this zone to converge toward the inner fetal nuclei in the periphery. This differs from the peripheral divergence seen in all of the peripheral areas (equatorially) of the outer zones of discontinuity. It should be remembered that the greatest sagittal growth of the lens occurs during embryonic and fetal life. The lens is somewhat spheroid. Although the lens fibers continue to develop after birth, the shape of the lens flattens. This means that the peripheral parts (between the zones of discontinuity) thicken relatively more than the axial regions, resulting in peripheral divergence of the zones of discontinuity. According to Vogt, the peripheral divergence results from a thinning (sagittally) of the individual lens fibers as their ends abut upon the suture, especially as the sutures (cortical and adult nucleus) grow in length. So that the sagittal thickness of each fiber, except for the fetal nuclei, is less in the axial regions than in the periphery. This state of affairs is abetted by the tension of the zonule.

ing, in contrast to the lower arms the ends of which practically always branch in a dichotomous way. As in the case of other sutures a slight deflection in the course of the main arm precedes the subbranching. Histologically, Rabl has shown in pigs that the posterior Λ -suture develops earlier than the anterior. According to Vogt this is probably also true in humans, as indicated at birth when the posterior Y is already further developed than the anterior. His measurements of the length of the fetal suture legs (made in fetuses of from 6 to 8 months) were approximately 1.25 mm. to 1.5 mm. for those of the posterior Λ up to the sites of subbranching and from 1.5 to 1.75 mm. for the lower leg of the anterior Y.

The posterior Λ , situated directly in the axis of the lens, aids in locating the posterior pole of the lens when looking for remains of the arteria hyaloidea propria, since the polar area of the posterior capsule lies behind and nasal to it.

THE POSTERIOR OUTER FETAL (EMBRYONAL-VOGT) STRIPE

This stripe, which corresponds to the anterior outer fetal stripe, likewise may not always be sharply delineated (Fig. 364). Because of the greater curvature of all the posterior zones of discontinuity as compared to the anterior, the radius of curvature of the posterior outer fetal stripe is less than that of the corresponding anterior one. With age, sclerosis of the central and adult nuclei also tends to diminish its visibility. Added to this is the commonly seen increase in yellowness of the posterior parts (posterior cortex and posterior adult nucleus).

THE POSTERIOR ADULT NUCLEAR STRIPE *

The stripe of the posterior adult nucleus is the posterior counterpart of the anterior adult nuclear stripe. This zone, which also is the thinnest in the axial region, broadens out somewhat as it approaches the periphery and shows the same characteristic peripheral divergence. (See Figs. 362, 363.) In other words, the distance be-

* Corresponding to those in the anterior cortex, fainter accessory bands may usually be seen in the posterior cortex. However, these will hardly be confused with the more distinct and reflecting stripe of the posterior adult nucleus.

tween the stripe of the posterior adult nucleus and the posterior line of disjunction is greater peripherally than axially. The posterior surface of the adult nucleus likewise becomes more marked with age and shows the same type of relief as the anterior but in reverse, so that a similar structure that appears as a prominence on the surface of the anterior adult nucleus will appear as a depression on the surface of the posterior and correspondingly a depression anteriorly will appear as a prominence posteriorly. The same fiber design and the same variations occur as in the appearance of the relief of the anterior adult nucleus surface. The zone of the posterior adult nucleus should first be localized (just behind the posterior Λ) by optic section axially, where the markings are most pronounced. Then, in order to get more surface area of reflection the pre- or post-focal part of the wide beam should be employed. To obtain a larger (specular) reflecting zone a change in angulation between illumination and observation may be indicated. As previously described, the mirror zone of the posterior adult nucleus surface, although somewhat smaller than the anterior, is more orange in color, especially in older individuals (Plate LVIII, figs. 2, 4). This is even true when it is compared to the shagreen area of the posterior lens capsule (page 974). Sharp focusing may in some cases disclose a delicate fiber design, but ordinarily it appears, depending on the width of the slit opening, as a small rectangular area of illumination covering only a small part (in height) of the posterior adult nucleus stripe. As the beam is diffused (pre- or postfocal illumination) the lateral edges become less sharp and gradually fade into the surrounding non-specular areas.

THE POSTERIOR STRIPE OF DISJUNCTION

Sharp focusing of the posterior capsule stripe with the narrow beam will disclose a double stripe which, except for its greater curvature, seems to differ in no way from that of the anterior double stripes. The one just in front of the posterior capsule stripe is the posterior stripe of disjunction (Figs. 351, 364). That this zone of discontinuity does not represent the inner limit of the epithelial

layer is proved by the absence of epithelium histologically in this posterior zone. It probably represents a well-defined stratum in the subcapsular cortex in which, owing to a physico-chemical alteration of the fibers, a change in index of refraction occurs which is visible only in optic section. This stripe also reveals the typical peripheral divergence equatorially; it appears homogeneous and has no apparent surface markings.

THE POSTERIOR CAPSULE STRIPE

This stripe delimits the posterior surface of the lens and, depending on the width of the focused beam, represents an illuminated section of the posterior capsule (Plate LVIII, fig. 2). Ordinarily it appears as a curved bluish-gray stripe (concave anteriorly), its curvature corresponding to that of the posterior capsule. This surface can be focused in two ways, either by focusing directly along the path of the beam to the place where the lens relucency ceases and the fibrillar structure of the trembling vitreous framework comes into view or by focusing on the zone of specular reflection of the posterior capsule. As previously mentioned because of the thickness of the lens it is impossible to observe its entire sagittal thickness at one time in the focal part of the beam even with the 10-mm. illuminating lens or the Poser diaphragms. (See Vol. I, pages 48, 77.) When the focal part of the beam passes through the anterior parts of the lens, the posterior parts are illuminated by postfocal or somewhat divergent rays and the zones of discontinuity are no longer sharp (Plate LVIII, figs. 1, 2). Conversely, when the posterior parts of the lens are traversed by the focal part of the beam, the anterior illuminated section is seen in the prefocal part of the beam. After the posterior capsule stripe is sharply focused, lateral movement of the beam (until the angle of observation equals that of illumination) will reveal the zone of specular reflection (posterior shagreen) (Plate LVIII, fig. 2). If one wishes to obtain specular reflection from a point more nasally, then the patient is instructed to direct his gaze in that direction, or temporally for the mirror region of the temporal parts of the capsule. Alternately, if the catoptric image

of the illumination lens* of the posterior capsule is focused and it appears in the anterior parts of the lens and holding this image in view, deeper focusing of the microscope will suddenly reveal the shagreen of the posterior lens capsule when its point of focus corresponds to the posterior capsular plane. In this manner a shagreen picture is obtained which, although smaller, is even brighter than that of the anterior mirror region. The design of the posterior shagreen† is much finer than that of the anterior capsule, but it shows in addition reflections from the posterior subcapsular fibers as they radiate toward the sutures. The posterior cortical sutures appear as dark lines crossing the mirror reflex and are best seen just outside the axial region. As already described (page 972) in the discussion of the origin of the shagreen of the anterior lens capsule, the simultaneous appearance of the fiber design and suture lines in the shagreen does not necessarily indicate that they contribute to its formation but rather that the coincidence results from a superposition of the separate optical phenomena, *viz.*, separate specular reflection on the one hand from the lens fibers and on the other hand from the posterior capsule. (See also Morgagnian cataract, etc.) Optically, just as in all other reflecting surfaces, defects (depressions or elevations) are nonreflecting in the plane of specular reflection and appear as dark spots in a luminous background. (See Vol. I, page 93.) Normally in the axial region the mirror zone may show dark lines or dots resulting from interference caused by residua of the fetal tunica vasculosa. In focal light these remains (page 1020) stand out as whitish reflecting structures. The presence of these remains is practically universal; they will be taken up in detail under

* In the Vogt method of illumination, the spiral of the wires of the lamp (source of illumination) will be seen and in the Koeppe-Poser model the ground glass surfaces of the condensing lens system screens out the image of the wires and all that is seen is a rectangular figure of illumination corresponding to, but naturally much smaller than, the size and shape of the illuminating lens itself.

† The absence of any epithelial layer in front of the posterior capsule precludes the possibility that the design in the mirror reflex is caused by these cells. Koby states: "When a large vacuole exists at the posterior surface of the lens, we have sometimes been able to show that the convex part of the vacuole turned toward the observer, exhibits some shagreen. This observation demonstrates clearly that the lens capsule does not play any important part in the production of the appearance of shagreen." However, this conclusion is not shared by other workers, as it is now generally believed that the shagreen is an optical manifestation of the posterior capsule.

the heading of congenital anomalies. In the same way any pathologic feature also interrupts the specularly reflected rays from a given area. Whereas in the mirror zone these defects appear dark, in direct focal illumination their color is evident as, e.g., white cellular conglomerations, reddish or yellowish blood pigment and brown uveal pigment. Normally the shagreen varies little in appearance from case to case although its design tends to get more marked with age (shrinkage phenomenon). With complete opacification of the sub-capsular cortex, the shagreen gradually becomes indistinct.

THE CORTEX AND THE ADULT NUCLEUS

For our purposes, the cortex includes that part of the lens situated anteriorly and posteriorly between the capsule and the anterior and posterior stripes of the adult nucleus; while the adult nucleus is bounded outwardly by the anterior and posterior stripes of the adult nucleus and inwardly by the anterior and posterior outer stripes of the fetal nucleus.* The cortex, formed by the youngest fibers, is softer and more elastic than the substance contained within the deeper adult nucleus. As already mentioned, although the cortex continues to grow by the apposition of new fibers and as a result gets thicker with age, actually the total sagittal thickness of the lens changes but little. This retention of total lens flatness results not only from the sclerosing and shrinking process within the nucleus but also from the fact that as the cortical sutures of the lens divide

* At the time of an extracapsular cataract extraction the hard nucleus may appear quite large, even larger than one would expect from the depth of the cortex seen in older people with the biomicroscope. In these cases very little if any cortex remains behind. In all probability the sclerosing process extends into the deep cortex or outside the layer which, from the standpoint of biomicroscopy, we consider to be the limiting surface of the nucleus. This would indicate that the process of sclerosis is in no way related to the increase in index of refraction which optically governs the zones of discontinuity. Except for this instance, the cortex, particularly its middle and anterior portions (subcapsular), reacts in an entirely different way from the nucleus to the effects of noxious influences. The harder nucleus merely becomes diffusely cloudy and more relucant, its fibers do not undergo degeneration and solution, with resulting lamellary separation, formation of water-clefts, spokes, vacuoles, etc.—eventually forming opaque masses as is the case in the cortex. The difference in constitution between the central parts of the lens and the cortex is even exemplified by the behavior of hereditary postnatal opacities. Some of these are found predominantly in the axial regions of the central nuclei. Some others, also genetically determined, are chiefly formed in the periphery at the level of the surface of the adult nucleus or deep in the cortex (e.g., coronary opacities). The former are uniformly stationary whereas the latter have progressive tendencies.

and elongate, the ends of the fibers abutting them of necessity become flatter anteriop dorsally in the axial regions.

As the beam passes through the cortical thickness it appears slightly relucient and gray-blue in color. Occasionally in adults, one or more faint concentric accessory stripes of discontinuity may be seen within it. The complicated suture systems within the cortex are usually not to be seen in the young, except in the mirror regions. In the aged, and especially as the cortex becomes more relucient due to appearance of the so-called presenile cataractous changes (water slits and lamellary separation), the elaborate branching of the suture systems becomes more apparent (Figs. 360, 397). However, even in the young we may find some indications of the suture system of the adult nucleus surface if some relief is present. In adults and in the aged this relief becomes increasingly visible (page 994). Although the cortical sutures and especially those of the adult nucleus appear to lie in one plane, in reality they have a sagittal thickness, extending through the substance from before backward. Because of internal reflection, the biomicroscope does not permit its observation in a uniform or continuous manner. It can only be seen in macerated lens preparations. In such preparations maceration causes the lens to open in sectors owing to rupture of the sutures, and it has been found that the sagittal (depth) extension of the sutures gets greater as the axial regions are approached. However, this splitting does not go through the entire nucleus (Fig. 367). The nucleus splits into two halves in the region of the dark interval. Clinically this separation is seen in cases in which at operation a double nucleus is found. Topographically, in macerated preparations after rupture of the sutures, Vogt likened the intervening sectors to the sections of an orange; however, the sectors are actually made up of concentric layers more nearly resembling those of an onion. Biomicroscopically, the sutures and the zones of discontinuity graphically suggest this type of lens structure. As each new concentric layer develops below the capsule, the older one internal to it is pressed inward. This is clinically illustrated by the subcapsular opacities appearing after an acute attack of glaucoma or following trauma

which, with time, are found to recede deeper and deeper from the capsule. In addition to the well-known radially directed fibers, there are also concentric fibers forming concentric layers apparently inde-



FIG. 367



FIG. 368

FIG. 367. Preparation showing splitting of sutures and concentric layer formation. (After Vogt.)

FIG. 368. Punctate opacities in the anterior cortex.

pendent of the others. The radiating fibers were first seen by Rabl in his histologic preparations. He could not demonstrate the concentric lamellae. The anatomic fact of the difference in the direction of these fibers is interesting to us because it explains the basis for the variations in direction of the white lines seen in lamellary separation of the cortex and also concentric arrangement of opacities seen in certain types of cataract, e.g., coronary (page 1095).

Biomicroscopically, it is apparent that the suture systems become extended and more branched the closer they are to the capsule. Again, the arrangement anatomically governs the maintenance of the flat lens in spite of the continual growth of new fibers in the cortex, thus insuring the lens its ability to act in conjunction with the pull and relaxation of the zonule in the process of accommoda-

tion (see below). In addition to this the sutures probably play an important role not only in lens metabolism but also pathologically as a "locus minoris resistentiae." The ease with which they are separated by fluid forms the basis of the water-clefts or slits. In siderosis and chalcosis lentis the metallic deposits are first found in the sutural areas.

Frequently in adults small isolated white dots are seen in the peripheral cortex. Since, ordinarily, they do not tend to increase either in number or size and apparently have no connection with the onset of presenile or senile cataractous change, they are considered physiologic (Fig. 368).

The Increasing Yellowish Coloration. Another interesting change, especially in the posterior parts of the lens (cortex and nucleus) is the increasing appearance of yellowness of these parts with age (Plate LXX, fig. 5). Although this is a senile phenomenon, like other senile changes, it may not parallel age in every case, e.g., graying of the hair. Thus, there is great variation among individuals as regards the time of its development (possibly of genetic determination). As a matter of fact this is also seen in the same individual, where the increase of yellowness in the deeper layers may appear in one eye only or in one eye some time before it develops in the fellow eye. Unquestionably in addition to senescent changes, pathologic states of the lens, especially those in the posterior cortex induced by intra-ocular disease, e.g., glaucoma,* predispose to this color change. This change is thought by some observers to be due to an increase in the amount of tyrosine, an amino acid (the precursor of melanin). Others explain its presence optically as a manifestation of fluorescence. When light passes through the human lens even in the very young,† ultraviolet, violet and blue rays (of short wave length) are absorbed to a certain degree. One of the effects of this absorption is the production of fluorescence. With age this absorption increases and as a consequence the deeper portions of the lens have a yellow

* This has been noted particularly after antiglaucoma decompression and filtering operations.

† According to Vogt, even the lens of an infant has a slightly yellowish tinge when compared to the colorless lens of a calf.

to reddish coloration. In nuclear cataract when this process is extreme, *cataracta brunescens* (brown) or *nigra* (reddish-black) results (Plate LXX, figs. 1, 2, 3, 4). Optically the effect of this increase in yellowness in the posterior cortex and nucleus can be simulated in a normal person by wearing a yellow lens. To some degree this prevents the passage of the shorter rays through the lens and results in a relative form of "blue" blindness. This interference with recognition of delicate shades of blue has been noted in the work of aged painters. For example, in his later years Tintoretto's blues became more brilliant. The absence of this yellow filter in aphakia accounts for the complaint of "blue vision" in patients after operation for removal of the lens.* As this color-absorption phenomenon progresses, it not only affects blue but tends to pass over to green and even to yellow (yellow-blindness of Vogt). In the latter case, the yellower the light (rays of long wave length), the more difficult will it be to differentiate between shades of yellow. Vogt refers to the classical example of such yellow-blindness in the masking of the yellow color of the macula lutea when viewed in the ordinary light of the ophthalmoscope.

With continuing senescence, the yellowish color is most intense in the posterior cortex and gradually fades out in the region of the posterior adult nucleus. In extreme age this change in color advances diffusely, gradually becoming less intense and diluted in appearance anteriorly. One is reminded of the effect obtained in further dilution of an iodine solution, which is reddish to dark-brown in concentration and pale-yellowish with dilution. Optically, as the focal beam passes through the lens, the posterior cortex is seen through all layers in front of it. In other words the light is seen through all layers in front of it. Correspondingly, the light, as seen by the observer, is filtered twice—once as it enters and again as it emerges. This may be a factor in the unusual color of the mirror regions of the posterior adult nucleus (page 974). Bucklers³⁷⁴ has shown

According to Wald, aphakic eyes are 1000 per cent more sensitive to ultraviolet light than phakic eyes. His experimental work disclosed the interesting fact that persons with aphakia can read a chart illuminated by "black light" otherwise invisible to phakic individuals.

some beautiful illustrations demonstrating spectroscopically the absorption of colors as light is passed through yellow filters.

RELATIVE PROGRESSIVE CHANGES IN THICKNESS BETWEEN
THE CORTEX AND ADULT NUCLEUS

In order to confirm the fact that the cortex increases in thickness progressively with age and that paralleling this there is a relative decrease in thickness of the nucleus in relation to the total lens thickness, Vogt⁶⁵³ made elaborate and extensive lens biomicroscopic measurements on 19 persons varying in age from 12 years to 77 years. He made his measurements with an ocular micrometer, considering the total thickness of the lens as 1 (Fig. 369A, B). His results were percentage figures. Analyzing them (anterior and posterior cortices combined) as compared to that of the adult nucleus, Koby (after Gallati⁴⁴¹) prepared the following table:

TABLE XVII
RELATION BETWEEN AGE AND THICKNESS OF
CORTEX AND NUCLEUS *

AGE OF SUBJECT (YEARS)	CORTEX (PER CENT INCREASE)	NUCLEUS (PER CENT DECREASE)
16 — 19	0.178	0.822
20 — 29	0.207	0.793
30 — 39	0.256	0.744
40 — 49	0.287	0.713
50 — 59	0.309	0.691
60 — 69	0.319	0.681
70 — 81	0.329	0.675

Vogt found that the axial cortical thickness (anterior and posterior) at the end of the second decade and during the third, comprises a much smaller part of the total lens thickness than in the later decades, and that the axial cortical thickness steadily increases in extreme age. At the same time, the nuclear thickness becomes less in relation to the total lens thickness, being more than four-fifths in

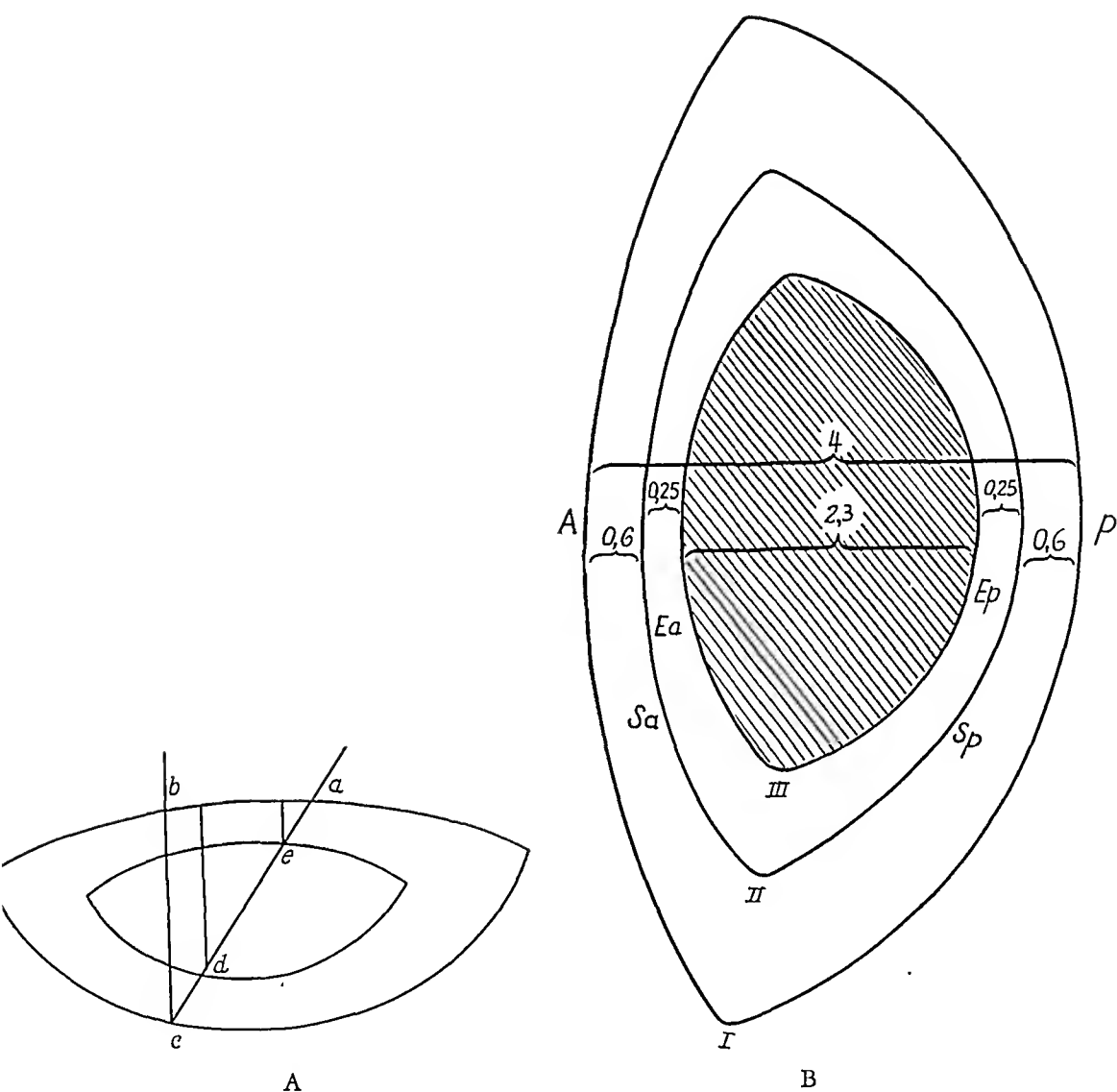


FIG. 369. A. Diagram illustrating the method of measuring the distance between zones of discontinuity. An illuminating lens of long focus is used: $a-c$, direction of the light beam; $a-b$, frontal plane on which segment (ac, ed, dc) in the optic section are projected; $b-c$, direction of observation. B. Diagram of distances of the more important zones of discontinuity in the adult lens. The distances do not vary between the age of 20 and 40. A , anterior capsule; P , posterior capsule, Sa , surface of the anterior adult nucleus; Sp , surface of the posterior adult nucleus; Ea , surface of the anterior outer embryonal nucleus (fetal nucleus); Ep , surface of the posterior outer embryonal nucleus (fetal nucleus). The total estimated thickness is 4 mm. Each cortex (axial) is 0.6 mm.; adult nucleus is 2.8 mm.; embryonal nucleus (fetal and embryonal) is 2.3 mm. The thickness of the cortex (axial) in the young adult is one-seventh of the total lens thickness, while in older persons it is one-sixth.

the second decade as compared to two-thirds in the seventh decade. He further showed that the increase in cortical thickness is not only relative but absolute by comparison with the thickness of the cornea (which is relatively constant). In the young, the thickness of the cortex is somewhat less than that of the cornea whereas in adults its thickness is two or three times that of the cornea.

RELATIONSHIP OF THE MORPHOLOGY OF THE LENS TO ITS FUNCTION OF ACCOMMODATION

In spite of the relative increase in thickness of the cortex during life, seen in the axial region, the lens maintains its flat shape. As already indicated, this is accomplished by the development of a complicated system of sutures. As the sutures elongate and branch, the abutting fiber ends broaden out; this can only be accomplished by a process of sagittal flattening of the fibers (anteroposterior). That this does not occur in the periphery (equatorially) is evidenced by the peripheral divergence of the zones of discontinuity in this region, signifying an increase in the total sagittal thickness of the layers between the stripes of discontinuity. The only exception to this is seen in the inner fetal nuclei, which are thicker axially than equatorially and in this sense retain their original more or less spherical (coffee bean) shape. Comparative anatomy illustrates the increasing tendency toward a flat lens (anteroposteriorly) as the phylogenetic scale ascends. In the lower series, at rest, in contrast to that of man, the soft homogeneous lens is spheroidal and accommodation occurs crudely by changes in position of the lens or by circular compression.* In lower mammals (e. g., rodents) the lenses are spheroidal and have only a rudimentary suture system (i. e., a single line) while in higher mammals, a simple trichotomous suture is found, with the zones of discontinuity almost parallel and having little or no peripheral divergence. In the human newborn, the lens is not dissimilar to that seen in lower mammals. Its shape is somewhat spherical and just below the capsule are the simple three-

* This process (e.g., in birds) probably gives a greater range of accommodation at the expense of clearness, in contrast to the opposite effect arising from simple relaxation, as in mammals.

branched Y-sutures. From then on the lens continues to grow by the interposition of new fibers below the capsule but at the same time, it becomes flatter anteroposteriorly. Undoubtedly the pull of the zonule is an important factor in this process. Biomicroscopically, observation of the zones of discontinuity and their peripheral divergence permits the ontologic as well as the phylogenetic development of an organ to be seen in the living for the first time. With relaxation of the zonule, during accommodation the anterior curvature of the lens increases (the radius of curvature according to Gullstrand is reduced from 10 to 5 mm. and the anterior pole of the lens advances by 0.3 to 0.4 mm.) while the posterior curvature changes but little. In the absence of flattening no accommodation could occur. This is exemplified in spherophakia owing to the deficiency of zonular tension, the elastic capsule containing the soft inelastic lens fibers tends to take a spheroidal shape, the lens assuming a condition of permanent and extreme accommodation. Evidently as the zonule relaxes, something occurs in the inelastic lens fibers to permit a thickening in the anterior axial parts of the lens. Gullstrand theorized that, owing to the speed of accommodation, fluid could not pass back and forth through the walls of the lens fibers and, owing to their inelasticity, their volume remains unchanged during the period when the zonule is relaxed. Hence he argued that the change in shape of the lens resulted from a change in relation of the radial fibers to one another within the lens (an intracapsular mechanism of accommodation).^{*} Contrary to this idea of shifting (and even interposition) of the fibers, Vogt believed — on a sounder morphologic basis — that the fibers change their shape, and that during accommodation a shifting of their contents may occur in the direction of the axial region, with a resultant increase of cortical thickness in this area. As an analogy he cited the thickening in fibers of striated muscle. The fact that peripheral divergence decreases during the act of accommodation bears out his hypothesis that an axialward shifting of the lens fiber contents occurs.

^{*} Despite the fact that the fibers are fixed at both ends and probably have a cement substance between them, Gullstrand believed that during accommodation the fibers in the plane of the equator change their position in the direction of the lens axis.

The exact role in accommodation exerted by the so-called "sclerosis" or hardening of the nucleus,* an apparent physiologic manifestation of adult life, is not entirely understood. Although not sufficient to affect vision, comparison of the central parts of the lens in the young with those of normal adults reveals in the latter a slight increase in relucency and opalescence.† Undoubtedly this increasing hardness in the nongrowing part of the lens acts to stiffen it, a condition that militates against the act of accommodation, save for the fact that this condensing process gives more room for the growth of the softer cortex. Evidently, at least until presbyopia sets in, the effect of the process of condensation of the nucleus in retarding accommodation is compensated for by the progressive flattening of the lens. As Vogt has pointed out, although sclerosis lowers the range of accommodation, the hardened nucleus is important for the maintenance of transparency of a large part of the lens during age. Pathologically, it resists the effects of noxious agents much better than the younger cortex to which falls the mechanical task of intracapsular deformation (accommodation). The sclerosed nucleus reacts to changes by becoming uniformly cloudy (nuclear cataract), a condition which as a rule is not as serious to vision as cortical disintegration and degradation characterized by the formation of completely opaque material.

MODIFICATIONS ASSOCIATED WITH AGING

As already mentioned, the accident of birth, onset of puberty, or approach to adult life does not interfere with or cause any sudden visible alterations in the ordinary predetermined life cycle of the lens. The physiologic aging of the lens proceeds in an orderly way, conditioned by the special racial as well as by individual genetically determined forces. Certain of the so-called "senile" changes may

* The term "sclerosis" is employed to designate a condensation or hardening (lack of elasticity) rather than opacification. When the nucleus becomes opaque in the sense of a cataract, it is known as a "nuclear cataract."

† Actually in the very young the narrow beam will show that the relucency of the cortex is somewhat greater than that of the nucleus. This difference may result from the greater anatomic and optic homogeneity of the nucleus.

appear earlier in life (abiotrophy) and even though they are hereditary they may be organ specific and not general. Examples of this kind of change are met with every day, e.g., premature graying of the hair, arcus senilis, pinguecula, and presenile and typical senile cataractous changes. The list of examples could be continued indefinitely and could include innumerable illustrations indicating hereditary tendencies toward premature aging apparently localized to certain organs or systems of organs (skin, blood vessels, etc.). Some of these changes are harmless, others interfere with vital processes and may lead to premature extinction of the individual. In Vogt's studies of identical twins, the symmetrical and uniform appearance of certain senile changes in each pair seems to provide incontrovertible evidence that these alterations are genetically determined. However, the effects of exogenous factors (e.g., nutrition) cannot be entirely denied, but in some cases even these may be genetically determined (e.g., certain endocrine disturbances). The ramifications of this subject would take us too far afield. The following list includes the senile changes which, from the standpoint of biomicroscopy, do not necessarily lead to a degree of opacification warranting the term "cataract." (Alterations, such as lamellary separation, widening of the sutures, water-clefts and types of "presenile" opacities more intimately associated with cataract, will be dealt with in a special chapter.)

1. Certain capsular (graining, spherules of the shagreen) and subcapsular changes
2. Increase in cortical thickness and its association to lens flatness
3. Increase in visibility of zones of discontinuity and suture systems of the cortex and adult nucleus
4. Increase in internal dispersion or optic density of the cortex and the nuclei (as seen in optic section)
5. Formation of the adult nuclear relief
6. Increasing yellowish coloration in the posterior cortex and posterior adult nucleus

PHYSIOLOGIC REMAINS OF THE TUNICA VASCULOSA LENTIS

These rests are included in this chapter on normal lens findings rather than under the heading of congenital anomalies because biomicroscopically, evidences of them are found in varying degrees in the posterior lens capsule of every eye. In a sense their presence cannot be considered any more anomalous than the finding of a fetal nucleus in every normal lens. With the exception of rare cases of extensive pupillary membranes these rests do not interfere with vision. The forms of residua of the tunical vasculosa lentis are enumerated in the following classification.

- I. Remains of the tunica vasculosa lentis:*

 - A. Tunica vasculosa anterior
 1. Pupillary threads and membranes
 2. Starlike (usually pigmented) figures
 3. Retro-iridal pigment lines and stripes
 - B. Tunica vasculosa posterior
 1. Whitish figures and strands
 2. Remains of the hyaloid artery (hyaloid corpuscle and vessel) strand

- II. Remains at place of insertion of hyaloid canal; arcuate line.

REMAINS OF THE TUNICA VASCULOSA LENTIS ANTERIOR

As discussed on page 959 the vessel net of the tunica vasculosa lentis anterior (pupillary membrane) is derived from the branches of the main trunk of the hyaloid artery (vasa hyaloidea propria and the capsulopupillary vessels).† Later branches from the ciliary vessels also supply the pupillary membrane. The double source of blood

* Retrolental fibroplasia in premature infants will be discussed in the chapter dealing with anomalies of the vitreous, page 1392.

† The lateral or capsulopupillary vessels are derived from the network forming the posterior portion of the tunica vasculosa lentis and extend to the equator of the lens where they anastomose with annular vessels. After the annular vessel is obliterated, the capsulopupillary vessels anastomose into the anterior arcades of the choroidal net. Buds from the annular vessels form the anterior tunica vasculosa lentis, the central part of which later is designated as the pupillary membrane. Eventually the ectodermal extension (future posterior surface of the iris) of the optic cup separates the primitive pupillary membrane from the capsule. As the iris develops branches from the ciliary vessels also enter to supply the pupillary membrane and the deeper iris layers.

supply to the pupillary membrane and the arrangements of this system helps to explain the origin of many of the rests as well as many other severe anomalies seen in postfetal life.

Persistent Pupillary Threads and Membranes. The pupillary threads and membranes (already referred to in Vol. I, pages 569-570 and Vol. II, page 755) frequently adhere to the anterior capsule of the lens. These membranes or threads are often connected with the iris frill (an important but not absolute differentiating point between them and inflammatory membranes, the latter more commonly being connected to the pupillary border), and either singly or branched extend over the pupil to be attached to the capsule. (See Vol. I, Plate XXXVIII, figs. 1, 2, 3, 4, 5, 6.) When they are delicate and white in color, which is usually the case in light colored irides, their attachment to the iris frill may be difficult to follow unless the pupil is dilated. In brown irides these threads tend to become pigmented and even the delicate ones may be brownish or at least have pigmented spots on them. At the point of attachment to the frill, they may thicken and fan out considerably forming well-developed brownish bandlike structures. Since the pupillary membrane is not pigmented in the fetus pigmentation obviously must occur later. Both filaments and stars (discussed later) are more prevalent in younger persons. Koby⁵⁰⁰ investigated 500 eyes for the presence of filamentous remains and epicapsular stars; they occurred at a more or less constant rate up to the fortieth year and from that age on, the frequency gradually diminished (although the stars tended to be the more persistent). In Koby's series, there were filaments or stars in 223 eyes examined, and both were absent in 277. Stars alone were found in 79 cases; stars and filaments together in 26. Although the filaments are quite elastic, it seems probable that pupillary activity ruptures them sooner or later; this would account for their infrequency in the older age groups.

Ordinarily, filaments and stars do not affect vision; however, on rare occasions a case will be seen in which heavy filaments are associated with a dense carpet of pigmented stars in the pupillary area. In one instance where this occurred bilaterally in a 40-year-old man

the vision could not be improved beyond 20/50 O.U. (See Vol. I, Plate XXXVIII, fig. 6.) In this case there were no pigment or other deposits on the posterior corneal surface. At times the attachment



FIG. 370. Small starlike pigment deposits on the anterior lens capsule.

of the filament to the capsule is marked by the presence of a small whitish opacity not unlike the hyaloid corpuscle of the posterior capsule. Whitish thickenings (sometimes pigmented) may also occur anywhere along the course of the filaments but are more pronounced where the filaments join the frill. As Vogt has pointed out such thickenings or condensations may correspond to the Wölfflin bodies seen in the periphery of normal irides (page 756). Persistent pupillary membranes also may be associated with anomalies such as colobomas and anterior polar cataracts, ectopia lentis, and spherophakia.

Starlike Figures. Numerous authors (Brückner,³⁷¹ Stahli, Kraupa,⁵¹¹ Koeppe,⁵⁰⁵ Koby,⁵⁰¹ and Vogt) have studied and described these figures on the anterior lens capsule in great detail (Fig. 370). Generally, they are located in the pupillary area but occasionally may be found peripherally also. Stars are found unilaterally or bilaterally. They usually are arranged in one or more groups, sometimes in the form of columns or chains; in the rare instances in which they are densely packed, they may form a mat (Vol. I, Plate XXXVIII, figs. 1, 2, 6). Their color usually corresponds to the color of the iris and may vary from dark brown to yellowish white. The characteristic shape of each star may be seen by high power. It may have three or four "points" with long tapering ends. The edges of the star between the points are concavely arched. Instead of being delicate and with three or four points, the mass may be clumped

with only a suggestion of one- or two-pointed processes or conversely the processes may be more numerous and spindly, resembling the spines of a burr—the longer spines tending to anastomose with

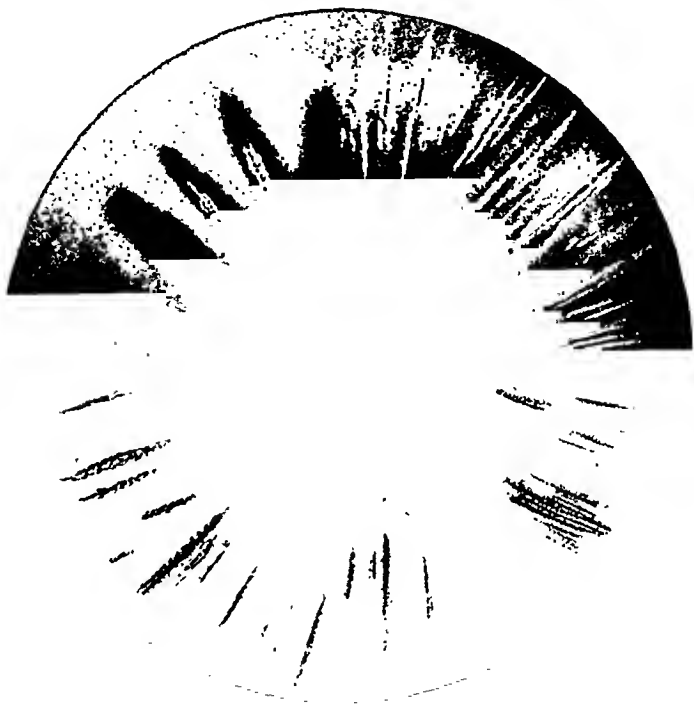


FIG. 371. Retro-iridal lines (nonpigmented).

neighboring ones. The stars vary in diameter from 20 to 60 μ . Koby has stressed the opinion that unlike the persistent threads, "the epicapsular stars are embryonic or fetal relics without any special relation to the pupillary membrane." Originally, it was held that all pigmented stars were fetal rests and that clumps of pigment were of inflammatory origin, but it is now known that starlike deposits may be found also following inflammation and trauma (Plate LXXIII, figs. 1, 2, 3, 4, 5, 6).

Retro-iridal Pigmented Lines and Stripes. These radiating grayish and pigmented lines and stripes on the peripheral parts of the anterior lens capsule were described by Vogt. Because of their delicacy and their peripheral location they have been overlooked. Wide dilatation of the pupil is necessary to see them. They are only visible in direct focal illumination with medium and high powers of magnification. They occur in normal eyes with no evidence of inflammation

and hence are not to be confused with the coarser pigmented deposits, which may also assume an irregular radial disposition, being derived from the furrow system of the posterior surface of the iris (page 846) following iritis. Vogt has termed these structures retro-iridal lines because he believed that they were rests derived from the retro-iridal part of the fetal anterior vascular tunic of the lens.* The appearance of these lines and stripes varies from case to case; some are composed of very short radiating lines of pigment, usually from 0.05 to 0.5 mm. in length, the longest reaching to 1.0 mm. With higher powers it will be seen that the lines are formed of fine pigment dots. In other cases, whitish lines or even broader pyramidal bands (apices pointing axially) may be found with or without occasional pigment grains. At times the axial ends of these whitish stripes and lines appear feathery (Fig. 371). In Vogt's series these lines and stripes were located more commonly on the nasal side and were stationary. I have seen them several times temporally situated as well, in older persons whose eyes showed no signs of inflammation. In one case especially there was senile atrophy of the pupillary margin. Since then I have observed these lines in younger individuals with no signs of iris atrophy. Recently Bellows reported three instances of the presence of these retro-iridal lines.

REMAINS OF THE TUNICA VASCULOSA LENTIS POSTERIOR

The biomicroscope reveals evidences of uniformly gray or whitish rests on the posterior lens capsule and in the anterior or hyaloid portions of the retrolental vitreous, in practically every normal eye. These structures, seen so universally with this method of examina-

* It was only after he found (in three cases) that some of these lines were in continuity with the remains (threads) of the pupillary membrane that Vogt was convinced of their origin. In one case, typical retro-iridal pigment lines extended axially in branching tracts of pigmented stars. Only the remains of the anterior vascular tunic are pigmented (the remains of the posterior tunic—on the posterior lens capsule—are always unpigmented except in inflammations). This may result from the fact that both parts of this anterior tunic, peripheral and central (pupillary membrane), receive blood from the ciliary vessels as well as from the posterior hyaloid system. The posterior tunic is derived solely from the branches of the hyaloid. Vogt has called the peripheral portion of the anterior tunic (the capsulo-pupillary part) the "retroiridal tunic" because of its location and differentiates its rests from those of the pupillary membrane by the fact that in the former they are seen as stretched radiating lines, whereas in the latter they are found in the form of threads and stars.

tion, are not seen with the ophthalmoscope, with the possible rare exception of the hyaloid corpuscle (the so-called "spurious posterior polar cataract");* when strongly developed it may cast a shadow



FIG 372. Remains of the tunica vasculosa lentis posterior and unusually prominent twisted remains of the hyaloid vessels.

posteriorly. With the arc lamp, the visibility of these structures is enhanced and in many cases numerous smaller ones, not seen by means of the ordinary nitra lamp, appear. Corresponding to the nasal position of the optic papilla, the location of these remains (tunica posterior and the insertion of the hyaloid canal) on the posterior lens capsule is not at the posterior lens pole but from 1.5 to 2 mm. nasally and slightly below as described previously anatomically by Seefelder.†⁶⁰⁶ Physiologically their presence at the pole,

* This structure was first seen ophthalmoscopically and correctly described by Mittendorf and has been referred to as "Mittendorf's dot."

† The central hyaloid vessels begin to regress in the eighth month of fetal life and lose their continuity with the vessels of the disk but still retain their connection to the lens. The distal or lens parts gradually coil up. At birth the main trunk may still extend backward but within the first few years it gradually takes a dependent position and is seen at times as a coiled cord attached to the lens with its free end hanging loosely.

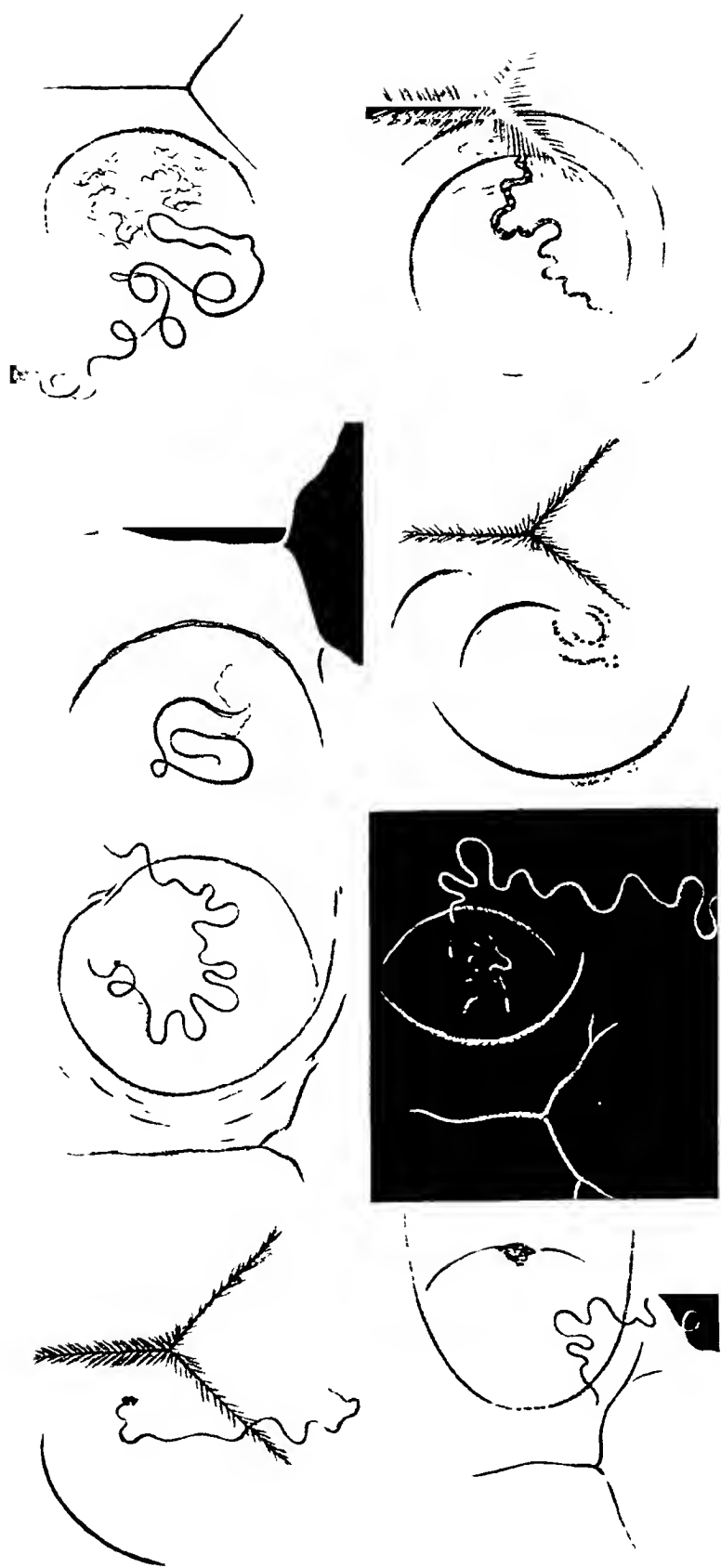


FIG. 373. Different forms (delicate white lines, nets, and whorls) of the remains of the tunica vasculosa lentis posterior. Note the arcuate line and hyaloid corpuscle.

especially if overdeveloped (from the failure of normal regression), would tend to interfere with central vision. Morphologically the shape and extent of these rests vary from case to case and even in

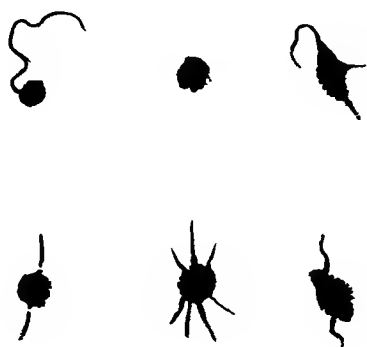


FIG. 374. Forms of hyaloid corpuscle.

each eye (although in the case of the latter they tend to be similar), but biomicroscopically some residua are always found. In order to see them it is best to localize the posterior Λ fetal suture because they lie behind it on the posterior capsule. If the beam (optic section) is directed from the temporal side, it will be seen that, as the angle between illumination and observation is narrowed, the distance between the posterior Λ -suture and the posterior capsule stripe seems to decrease. With the patient's gaze directed slightly nasally, it will be seen (when in sharp focus) that the rests lie on the posterior capsule nasally to the main sutural forking behind the Λ , or in other words usually just behind the upper subbranch of the lower nasal arm. If the mirror reflex comes into view at this point, it can be moved nasally and out of the way by having the patient direct his gaze still more nasalward. With experience the observation of this area becomes almost automatic.

In this area three distinct types of remains are found (with variation). However, they are not always seen fully developed in every eye, nor are all of them always present. (1) Delicate gray-white lines forming an irregular net or whorl. They may be fixed to the capsule throughout their extent or attached in parts; their ends may

float freely behind it. (2) The hyaloid corpuscle which may or may not have a twisted vessel cord or branches attached to it (partially free or attached). (3) The arcuate line, rarely a complete circle, commonly is seen as a crescentric figure on the posterior capsule nasally to or between the overlying posterior Λ -suture and the place of insertion of the vessels, its concavity facing the latter.

Whorl-like Remains. An exact description of the innumerable varieties of figures or designs formed by the whorl-like distribution of these delicate vessel-remains (which, according to Vogt measure from 0.03 to 0.06 mm. in thickness) would be too time consuming and laborious. The accompanying illustrations will give some of the forms ordinarily seen (Figs. 372, 373, 374). In some cases these structures are seen in the form of rings while in others they are curled.

The free-floating (nonfixed) curlicue or spiral line always seen first by beginners hangs vertically downward in the so-called "retrolental space" (anterior or hyaloid parts of the vitreous).^{*} Its attachment to one of the whorl-like convolutions of the fixed remains can usually be traced. With movements of the eye this nasally situated coiled line can be caused to snap like a whip, though more slowly. In some cases it may attain a length of several millimeters, extending deep or peripherally so that its terminal end will not be seen. Frequently small whitish spots or condensations occur along the course of these fibers. As a matter of fact, these spots are found on all the threadlike remains, especially on those in the anterior part of the vitreous. This curled structure (occurring in over 50 per cent of young people), which is much more commonly found than the corpuscles, becomes less frequent with age, either due to atrophy or as a result of being torn off. On the posterior lens capsule itself, it is rare to see any markings of rests except in the nasal area above described. However, in rare instances ring-shaped markings have been described on the posterior lens capsule in the far periphery. In one case (Hudson) the ring was found midway between the axis and the periphery. It was composed of a row of gray dots. In other cases

^{*} These, similar to the hyaloid vestiges seen deeper in this area not attached to the lens, are easily visualized because the surrounding darker or less reluctant vitreous offers better contrast than the normal opalescence of the posterior capsular stripe.

when located far in the periphery, it was formed of nonstellate amorphous brown pigment. According to Ida Mann this may result from the maintenance of contact between the tips of the ciliary processes and the lens for too long a period.

With the pupil widely dilated as described above, numerous branching fibers may be seen peripherally in the anterior vitreous (not connected to the lens) which evidently are derived from the vasa hyaloidea propria. Embryologically these peripheral branches of the artery are the first to show signs of regression. As early as the 60-mm. stage they tend to lose their connections to the main trunk of the hyaloid artery so that their free curled ends hang loosely in the vitreous, but distally they remain connected to the vessels on the posterior surface of the lens. The tendency to curl after separation is accounted for by their elasticity and, as noted above, is a feature of the large central branches and even of the main trunk.

The Hyaloid Corpuscle. These are densely white, more or less solid spherical structures, varying little in size and were formerly known as "cataracta spuria" (Fig. 374). They are located on the posterior capsule (where they frequently mark the attachment of the remains of the curled freely floating hyaloid vessel) or at times on the ends of floating remains behind it in the retrolental space or vitreous; those having an attachment to the posterior capsule are fixed while those attached to the mobile hyaloid vessel remnant move. The former are more common while the latter present a more striking picture. When very far back in the vitreous, they may even be visible ophthalmoscopically. In these cases they may only be discerned biomicroscopically when disturbed by ocular movements. Although, commonly, these opacities appear to be solid, spheroidal corpuscles, occasionally flattened discoidal bodies may be found. Vogt examined 82 children and found 7 who had corpuscles. In a study of 100 children (normal eyes) between the ages of 10 and 14 years, I found corpuscles in 11 cases. It may be considered that their frequency is of the order of 10 per cent in the young.

The Arcuate Line. In 1919 Vogt⁵⁴⁹ first described the so-called "arcuate line," which — according to its degree of arc incompletely or, less often, completely — surrounds the rests of the trunk and

the main central branches of the hyaloid vessels on the posterior lens capsule. Most often it appears as a whitish crescentric line, temporal to the place of insertion of the hyaloid vessels, or about half way between the area of insertion of the hyaloid vessels and the posterior pole of the lens (Fig. 373). It can be located easily by first focusing on the posterior λ . The arcuate line will then be seen on the posterior capsule directly behind it. Its convexity faces temporally toward the suture. In certain cases another more nasally located arc will be seen together with the usual one, with the ends of each intersecting due to a difference in curvature (Fig. 373). When both are present, their concavities face one another, their ends either crossing or, more often, fading out without touching each other. It is very rare indeed to find that the location of the arc varies either temporally (nearer to the lens pole) or nasally from its usual site. According to Vogt's measurements, the width of the arc varies between 0.02 to 0.06 mm., and in a few exceptional cases up to 0.15 mm. The radius of curvature of the arc is usually about 1 mm. The ends of the arcuate line behave in different ways but usually gradually fade out above and below, the vertical distance between the ends being from 1.5 to 2 mm. At times the two ends are stretched out so that a parabolic or hyperbolic figure is formed. In describing its optical properties while changing the direction of the light, Vogt found that it could be seen both by specular reflection as well as by direct focal illumination. He stated that in some cases, on moving the light (so that the angle of reflection did not equal the angle of incidence), he could cause it to disappear from view. This would suggest that the arcuate line protruded or was on a different level from that of the posterior capsule. In other cases, he found that changing the angle of the light did not cause the arcuate line to disappear. It is always seen in focal light, and hence its presence is not just a reflex resulting merely from a difference of level but rather is the expression of an opacity of the posterior lens capsule.*

* In his studies of the optical phenomena of the arcuate line Vogt found that in a certain proportion of the cases it was possible to cause the arcuate line to disappear by shifting the angle of illumination. This would seem to indicate that in these instances the arcuate line is an optical manifestation of regular reflection brought about not by the presence of an actual opacity but rather by a difference in level on the posterior lens capsule at this place.

Rarely, fine lines from the concave side may be seen running toward the hyaloid insertion. Also the area just temporal to the arcuate line may be traversed by delicate lines running concentrically with the arc. Vogt has also described in two cases out of a series of 200 that the arc instead of being a continuous line was made up of small radiating striae.

The exact derivation of the arcuate line is still uncertain. However, recent research (especially in the light of the proved existence of vestiges of the hyaloid canal in the retrolental regions of animals and also on the basis of comparative biomicroscopic findings), seems to indicate, according to Vogt, that the arcuate line which surrounds the area of insertion of the hyaloid vessels represents the place of insertion of the hyaloid canal. He points out, for instance, that the region, embracing the area of the insertion of the hyaloid vessels, and outlined by the arcuate line, in some persons appears darker than the outside surrounding regions.* On the other hand, it would appear that the anterior opening of Cloquet's canal should be larger than that indicated by the curvature of the arcuate line. Ida Mann suggested that the line marks the place where the early perilenticular tunica fibrosa was attached to the lens, i.e., before it was separated by the ingrowing vascular network derived from the hyaloid vessels. The diameter of this area at the end of the second month corresponds better to that of the arcuate line.

In other cases, notwithstanding, in changing the angle of illumination the arcuate line was constantly visible. It would seem therefore that a definite opacity was present and was visible at any angle of illumination by diffuse reflection.

Koby states: "A curious phenomenon in illumination may lead to error. It happens that the corneal reflex lights a part of the mount of the objectives, and this is reflected in the cornea in such a way that a semilunar image is formed in the deeper parts of the lens. This image so closely resembles the arcuate line that it may be mistaken for it. Their catoptric images are recognized from the fact that when the beam is moved slightly, two exactly similar images of the twin objectives are seen, and in the middle of each there is a luminous point arising from the reflections of the light at the surfaces of the objective themselves."

* This corresponds to the condition found in dogs and rabbits in which a dark (optically less relucant) area outlined anteriorly by the arcuate line extends backward into the vitreous. Within this darkened area the remains of the artery can be followed dorsalward. Also the gray lines concentric with the arc line occasionally seen in man are in these animals accentuated, giving this area a greater relucency. Vogt believes that this is the place where a reflection or reversal of direction of the membrane-like layer occurs to form the wall of the hyaloid canal. These conditions are better seen in animals because of the greater opacity of the vitreous and because of the lesser tendency of the vitreous to make oscillatory movements.

Chapter Twenty-Four

DEVELOPMENTAL LENS CHANGES

CONGENITAL ANOMALIES OF THE LENS

THE universally found rests of the vascular tunic thus far discussed have been considered "physiologic" in nature. Other anomalies of the lens (principally ectodermal) grossly deform the lens either in size, shape, or position or by the formation of opacities (cataracts) and interfere with function.*

In this discussion only the developmental (congenital and early acquired) lens changes will be considered. The progressive juvenile forms of coronary cataract (*cataracta cerulea* and *viridis*) and opacities associated with presenile and senile cataractous changes, which in turn may be considered also as developmental in the sense that they are probably genetically determined, will be considered under a special heading. It should be pointed out that except for their frequency and unless a thorough history and pedigree is obtainable, it may not always be possible to separate developmental cataract from morphologically similar instances of environmental causation. In other words it may well be that morphologically the pattern of lens opacities is governed more by temporal factors than by etiological ones. So that during the various periods of life lens changes,

* I should like to quote verbatim the opening paragraphs of Ida Mann's description of developmental cataracts, since it so beautifully summarizes the subject: "It is to be remembered that at no normal stage of development is the lens other than transparent. This property it shares with all other embryonic tissue (except the blood and the pigment cells) in early stages and it retains this property when, owing to more and more complicated differentiation, the other tissues have become opaque. Even in the lens the earliest formed part (the central dark interval) remains the most transparent (optically empty) throughout life, while the later formed secondary lens fibers show zones of increasing relucency and fluorescence until the most superficial fibers formed in extreme old age show opacities so often that these might almost be considered normal. Therefore all congenital lens opacities are pure aberrations and can never be arrests. Any opacity, whether total, or so small that it can only be seen by focal illumination with the slit lamp, is technically a cataract. Cataracts may be present at birth or may form during infancy or adolescence. In either case they are developmental and this term, rather than congenital, should be used for them, since in the lifelong growth of the lens fibers the moment of birth is only an accident. Even changes of commencing senility are not sharply marked off from growth processes."

whether of genetic or environmental origin, appear similar. An example of this is seen in zonular cataract (*cataracta parathyreoopriva*) and in diabetic cataract. Cases have been recently reported in Australia where rubella acquired by the mother during early pregnancy results in a congenital cataract in the offspring.* In toxoplasmosis also, congenital defects of the lens may result which morphologically may be very similar to developmental cataract of hereditary origin.

It is interesting to note that all the above (congenital, juvenile, presenile, and senile) developmental opacities seem to have a special predilection for certain specific zones in the lens. In other words, the different zones of the lens (cortex, adult nucleus, fetal nuclei) react at a certain time during the course of development to influences leading to cataract. However, these cataracts morphologically have different forms. It will be seen that not only do these opacities lie in the above-mentioned zones but in most instances, depending on the stage of lens development, they are found in specific parts of these zones and have individual distinguishing forms biomicroscopically. From this it could be concluded that during certain periods of life not only the main zones but even the subzones show specific changes which are governed by heredity, called "idiokinetic" changes. The same effect may result from the influence of noxious agents. In addition to the location-specificity of these opacities, there also is a relationship between morphology, location, and chronologic development. At the present time less is known about the chronology of development than the others.

The chronologic factor (time of inception), as regards the character of the anomaly, is more important whether the underlying causal factors be hereditary (genetic) or environmental (nutritional, toxic or mechanical). In either case when these forces act early, gross malformation or disturbance results. Later, after the pattern of the lens structure has been established, the resulting defects can only be an aberration, which may be simple, complicated, or may

* Most of these were total in character and were associated with microphthalmos. Goar and Potts reported seven cases all of which were accompanied by other defects, "usually congenital heart lesions, poorly developed musculature, and retarded mentality. The cataracts are typical in that the embryonal nucleus which develops soon after the fibers are laid down is affected." There were no indications of fetal iritis (*synechiae*).

form "lesions resembling the effect of disease." In environment defects, i.e., those resulting from pressure or toxins, gross deformities of the lens may result if their action is very early because (not unlike the retina) the tissue instead of reacting in the ordinary way tends to undergo changes comparable to necrosis. According to von Szily:* ⁶²⁷ "The early stages of idiokinetic malformations (hereditary) of the lens now permits of recognizing divergences from normal development especially in two directions. In one group we have to do with anomalies in the mutual delimitation of the two main sections of which the embryonal lens is composed, i.e., of epithelium and fiber components. The second group which indirectly also sets in a little later, represents disturbances in the formation of the sutures of the lens. Both processes lead to a more or less serious disturbance of the internal structure of the lens, and are mostly secondarily connected with disintegration of the lens sections affected." In environmental cataract (congenital or acquired) the alteration (e.g., as demonstrated by cataractous opacity) is primary, and any morphologic deformity occurring in consequence of it is secondary. However, clinically, as mentioned above, in the absence of a satisfactory pedigree recording repeated instances of the anomaly, differentiation of environmental and hereditary lenticular alterations may be uncertain.

At birth, the embryonal Y-sutures are situated closely behind the capsule. Although during the first years of life there are already indications of those portions of the lens that will later develop into the adult nucleus and cortex, it can be deduced that opacities occupying the central (embryonal and fetal parts) of the lens (e.g., anterior axial cataract, stellate cataract, polar cataract,† and cataracta centralis pulverulenta) are formed during fetal life, and zonular cataract probably develops just before or after birth.‡

The congenital opacities above-mentioned or those which develop just after birth are restricted to well-defined zones and do not

* The reader is advised to read the monograph published by von Szily in 1938.⁶²⁷

† Polar cataract (subcapsular) becomes separated from the nucleus by the ingrowth of secondary lens fibers.

‡ The larger size of this form of cataract—4 to 7 mm. in comparison with the size of the lens during fetal life—speaks for a later development.

progress; thus, they appear the same morphologically in age as in youth. We know little concerning the time of onset of the so-called "presenile" opacities (e.g., coronary and cerulean), which are somewhat progressive, although they seem to start at about the time of puberty. A great deal more detailed study — especially statistical — will be necessary before the time of onset of most of these opacities can be established.

Following is a classification of the various congenital anomalies of the lens:

- I. Congenital lens alterations due to gross changes in size, shape, or position
 - A. Congenital aphakia
 - B. Microphakia
 - C. Lenticonus, anterior and posterior
 - D. Lentiglobus
 - E. Coloboma lentis
 - F. Ectopia lentis and spherophakia
- II. Congenital developmental lens opacifications *
 - A. Polar and capsular cataract (anterior and posterior)
 - B. Opacities in the vicinity of the fetal Y-sutures
 1. Anterior axial embryonal (fetal) cataract (Vogt)
 2. Stellate suture cataract (anterior and posterior)
 - C. Zonular cataract (lamellar)
 - D. Cataracta centralis pulverulenta (Vogt)
 - E. Rare forms of congenital axial cataracts
 1. Cataracta pisciformis (Vogt)
 2. Coralliform cataract
 3. Axial fusiform cataract
 4. Spear cataract
 5. Floriform cataract

* It should be pointed out that most of these opacities are located predominantly in the axial regions. Hereditary opacities, e.g., coronary, cerulean and the so-called "presenile opacities" (see page 1095) which develop after puberty are located more peripherally (equatorial).

- F. Rare forms of congenital cataract affecting a greater portion of the lens
1. Disc-shaped or ring form cataract
 2. Congenital morgagnian cataract

GROSS CHANGES IN SIZE, SHAPE, OR POSITION OF THE LENS

Congenital Primary Aphakia. This very rare condition must be differentiated from congenital secondary aphakia in which partial or complete solution of the lens has occurred after it has already been formed. According to some investigators the former is only found associated with gross malformations, e.g., anophthalmia or severe microphthalmia. Whether or not it can occur in eyes not associated with other severe defects has not been established.

According to Vogt's cases congenital aphakia (hypoplasia lentis) occurred in association with microphthalmia, microcornea, nystagmus and central corneal opacities. The possibility of intrauterine infection with corneal ulceration must be considered, as has been shown by Seefelder.⁶⁰⁵ But Vogt questions this etiology in his 3 cases involving 6 eyes because of the symmetrical leukomata and their regular bandlike connection with the axial pupillary region, in that it could hardly be possible that in all 6 cases the lens degenerated in the same way. Anatomically v. Helmholtz showed that congenital aphakia occurs (having demonstrated it in a microphthalmic and hydrophthalmic eye). Following von Hess, Vogt believed that the disturbances in separation of the primary lens vesicle could be the cause of this anomaly. Early degeneration and aplasia of the lens could easily hinder the development of the cornea resulting in microcornea. In two of his cases ultraviolet light projected into the pupil gave no lens fluorescence. In one of his cases there was aplasia of the iris (only the mesodermic layers were present) with synechiae extending to the cornea and to the membrane in the pupillary periphery. In this case the lens was substituted by a band, consisting of scar tissue surrounded by a delicate film. After iridectomy the finely pigmented zonule fibers were seen overlying the delicate membrane. Optic section through the membrane revealed a double line, the

outer brown (zonule), and the inner or deeper gray (anterior limiting layer of the vitreous). These eyes are highly amblyopic and because of the nystagmus biomicroscopic examination is difficult.

Secondary aphakia occurs more frequently in otherwise normal eyes but more often it is associated with microcornea and other malformations. It likewise is thought to be due to primary failure of development in surface ectoderm. In these cases some vestige of degenerated lens matter and capsule (which may be wrinkled or shrunken and to which zonule fibers are attached) are always found. Secondary invasion by repair tissue and vascularization may further complicate the picture. Except for the microcornea or other defects, this picture is similar to cases occasionally seen in adults in whom, owing to long-standing disease the lens eventually becomes shrunken, calcareous and vascularized.

Microphakia (*spherophakia*, or lens "rotunda"). This anomaly is an example of sudden arrest in growth of the lens, which may occur without any apparent defects, except in the zonule.* Whether this anomaly is directly the result of a genetically determined inhibition of the lens anlage, loss of growth energy, or aplasia of the zonule is still unknown. According to Vogt, his biomicroscopic findings of torn and absent zonular fibers and such pathologic changes as pigment deposits indicate that in microphthalmos as well as in ectopia the underlying factor is not abnormally long zonular fibers (a view generally held) but rather the presence of rudimentary weak ones. This inherent weakness in the zonule is conducive to stretching and tearing of the brittle fibers. Since there is not sufficient power to exert tension upon the lens, normal development of the flattened shape is impeded. As a result the lens retains its spheric (embryonic) form. Because of the nature of the zonular changes further consideration of microphakia and *congenital ectopia* from the standpoint of biomicroscopy will be found in the chapter on the zonule (page 1341).

* It has been pointed out that in this condition the diameter of the cornea is normal or commonly is slightly enlarged. In cases of microcornea interference with the development of the lens is commonly seen, e.g., congenital cataract. It is still unknown whether genetically or mechanically there is any relationship between the size of the cornea and spherophakia.

Lentiglobus (*Lenticonus*). The terms "lentiglobus" and "lenticonus" describe an anomalous localized deformity of the curvature of the anterior or posterior lens surface. It is characterized by the presence of a conical or bullous projection or ectasia. When the protruberance is globular or hemispherical, it is known as "lentiglobus." Actually uncomplicated (true) protrusions are globular in shape while most of the complicated ones are conical.

Since the introduction of the biomicroscope, more of these cases have been brought to our attention, especially the posterior lentiglobus (anterior lentiglobus and lenticonus being more rare). According to von Szily, only those cases in which the protrusion occurs without rupture of the capsule should be considered as pure forms of lenticonus or lentiglobus.

Posterior Lentiglobus. In most instances globular projections, predominantly uniocular, have been found in the neighborhood of the posterior pole and particularly in the region of the arcuate line. Biomicroscopic descriptions of these cases have been given by many authors (Rydel, Vogt,⁶⁴⁵ Butler,³⁸¹ Whiting, March, Tyson⁶⁴³ and Pellathy⁵⁵⁹). In Butler's case a rosette-like opacity on the posterior surface of the infantile nucleus (future adult nucleus?) was connected to a smaller posterior disk by a stem resembling in shape a collar button. It was located at the posterior pole. He also reported a case of lenticonus internum or lenticonus perinuclearis posterior, apparently an abortive case in which the posterior capsule was unaffected. There was an unusual backward double bulge of the posterior adolescent nucleus. This appearance was caused by a concave depression in the axial part of the protrusion of nucleus. The area of the bulge was diffusely opaque. In March's case there was no opacification in the globular protrusion of the posterior capsule. Tyson described a case (unilateral) which was somewhat flattened or saucer-shaped (opaque). The diameter of the protrusion was about one-fifth that of the lens and the corrected vision was 6/30.

The degree of protrusion may be rudimentary or marked. In most cases, some degree of opacification is present, although cases have been described in which a hemispheric protuberance was found

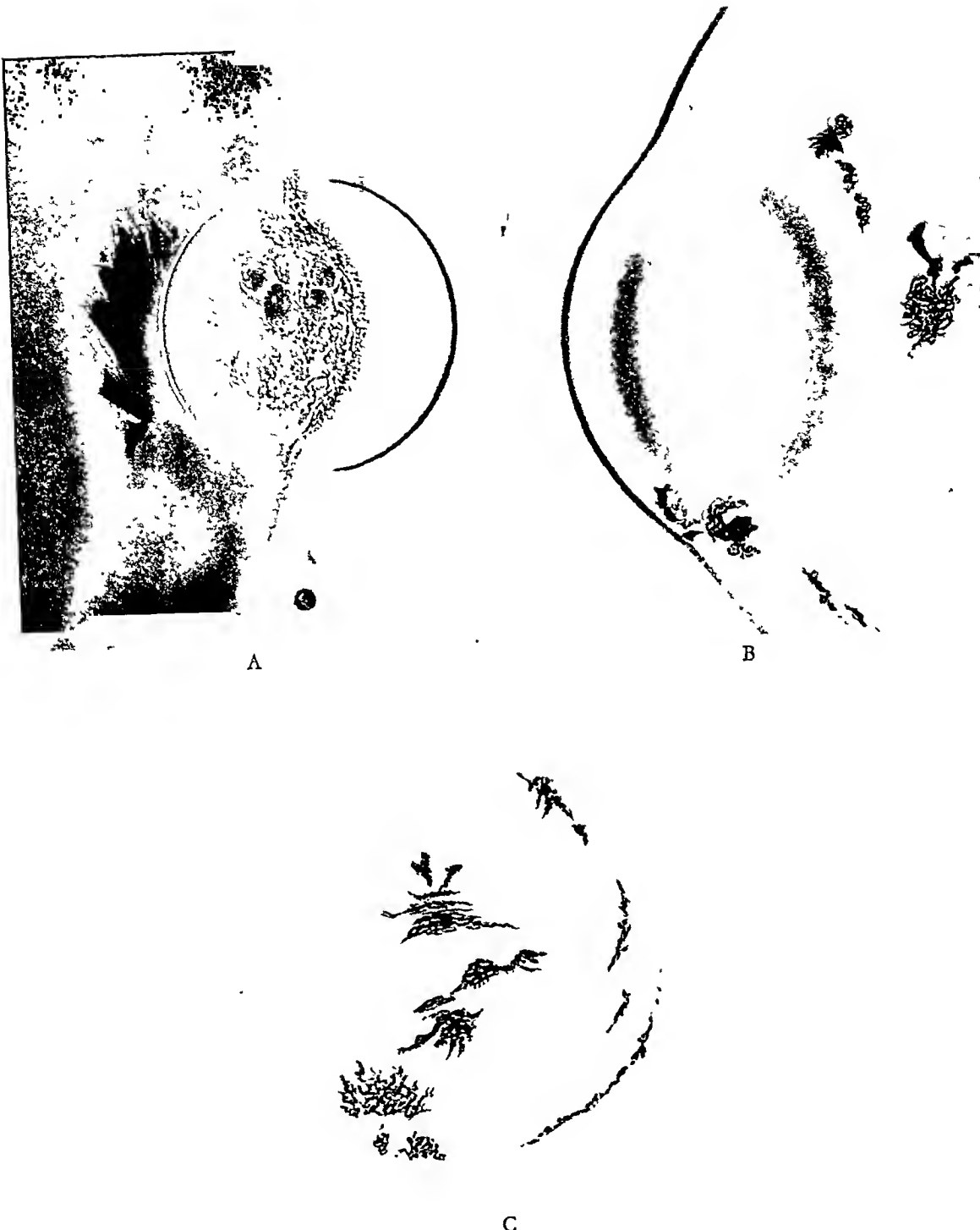


FIG. 375. A. Posterior lentiglobus. Note circular reflex, opacities and large water slit in posterior cortex; indentation of posterior adult nucleus. B. Lentiglobus posterior with dilacerated opacities. C. Same case as B. Detailed view of opacities.

to be transparent or clear in an otherwise normal lens. The location and extent of the opacity varies from case to case. An almost constant finding is a sharp and circularly rounded anterior margin of the conus. This results in a vivid ring reflex caused by a reflection from the capsule at the peripheral rim of the cone (Fig. 375 A). It is difficult to see this unless the pupil is dilated. The direction of the capsular curve suddenly becomes convex in a frontal direction before it begins to bulge dorsally. The sharp circular ring (reflex) results from specular reflection at this point and may appear doubled, if observed binocularly, since specularly reflected rays from one given point will fill only one eyepiece at a time. The reflex ring surrounding the protuberant area can at times be seen macroscopically or with the ophthalmoscope and when the conus is not clouded by opacities, it has been described as an oil droplet within the lens. It will appear reddish in the light reflected from the fundus. This form of retro-illumination can also be obtained with the biomicroscope if the beam is directed from the temporal side, and the conus can be viewed in the red light reflected from the nasal fundus. In addition Vogt has described scissors reflexes, i.e., two reflexes located behind the lenticonus which come from the opposite sides of the walls. These can be made to cross one another by moving the direction of the beam.

The presence of the ring reflex is diagnostic because it proves that a sudden change in capsular curvature is present. The sudden change in curvature is also directly visible in optic section (Fig. 375 A, B). In this way when the conus is clear or sufficiently translucent, it will be seen that the outer zones of discontinuity follow the abnormal curve. However, the inner fetal nucleus is never involved and will be found intact in its normal location. The opacity itself may outline the bottom of the cone posterior capsule and in some cases it will be seen that it extends laterally. The part outlining the walls of the protrusion may also differ in consistency and density, from the central part forming its base. Thus, when the opacity is viewed frontally in diffuse light it may appear as a small central circular area, surrounded concentrically by an opaque band. Also one may

find an opaque layer of opacity in optic section within the conus in front of those described above but located more centrally at the level of the anterior adult nuclear stripe (Fig. 375 A, B). The reduplication of layers reminds us of the structure of polar cataracts. Between these layers of opacity the cortex will show varying degrees of increased relucency. In other cases, a small opacity may lie within the conus without touching or involving the posterior capsule, but it may contact the posterior adult nuclear stripe or, in very young individuals, one of the stripes outside the inner fetal nucleus (which later are thought to fuse forming the adult nuclear stripe). According to the descriptions of cases in the literature most will fall under the heading of lentiglobus. It may be that lenticonus has a different pathogenesis than lentiglobus. Many suggestions by several writers have been proposed for the explanation of the pathogenesis of this anomaly. The fact that the fetal nucleus remains intact would indicate a late or postnatal development. Added to this is the frequent involvement of the zone of discontinuity which corresponds to the adult nuclear stripe formed just before or after birth. In most cases the site of the bulge is at the pole in the neighborhood of the arcuate line. This is temporal to the insertion of the hyaloid artery and might indicate that the conus does not result from direct traction of the hyaloid vessel on the posterior surface of the lens. In one of his cases, Vogt found the vestige of the hyaloid artery attached just at the nasal edge of the conus. (Normally the hyaloid remains are found within the arcuate line and not at its edge.) He theorized that if the development of the capsule in the region of the arcuate line (which normally according to Drusault shows a development different from the rest of the capsule since it attains its full thickness in the fourth fetal month) was faulty (weak or torn) it might give way to the intralenticular growth pressure and herniate. Bach,³⁵² basing his conclusion on a histologic demonstration of the attachment of a persistent hyaloid vessel to a posterior lenticonus, believed that the attachment caused a weakness or rupture of the posterior capsule with resulting herniation.

Von Szily (1928)⁶²⁶ found that detritus collects behind the lens

during its development and that this is later removed by the vasa hyaloidea. Failure of absorption of this material might also be a factor in interfering with the normal development of the capsule and hence might induce a herniation. Others have attributed it to an unequal traction of the zonule associated with inflammation or even accommodative effort. The cases studied were complicated and hence probably fall into the group classified as lenticonus rather than lentiglobus.

Anterior Lenticonus and Lentiglobus. Comparatively few cases (about 12) of this anomaly of the anterior face of the lens have been reported in the literature, and only in one or two instances have they been studied with the biomicroscope. Only about half of them were considered to be congenital. In one case cited by Bellows (Zavalia and Oliva⁶⁸³) there seemed to be a recessive hereditary transmission; two sons of a consanguinous marriage had anterior lenticonus (bilateral in one). Knienecker⁴⁹⁷ reported a case (also cited by Koby) of a bilateral anterior lentiglobus. The axial protrusions—which measure from 3 to 4 mm. in diameter and 2 mm. in height—were clear, and there were no other associated anomalies. The fetal nuclei were intact, and only the cortex was involved in the defect. In the region of the protrusion, retinoscopy revealed high myopia (−20. D). The peripheral unaffected parts were emmetropic. Feigenbaum⁴²⁶ also reported a case (unilateral) of anterior lentiglobus in a 12-year-old boy in which the fetal nucleus was involved. Many suggestions have been advanced to explain the pathogenesis of anterior lentiglobus. Among them are a faulty separation of the primary lens vesicle, a weakness of the anterior lens capsule with resulting herniation owing to the internal pressure of lens growth, and a lack of adequate or regular zonular tension because of an anomalous insertion or absence of the zonular fibers.* The fact that in some cases the protrusion developed later in life and was progressive was explained, according to Bellows, by Marsh⁵³⁶ and Feigenbaum⁴²⁶ as a result of accommodative strain in the presence of a congenitally weak capsule. In addition pathologically Seefelder and Wolfrum (1907)⁶⁸¹

* In the latter case should we not expect to find spherophakia or ectopia lentis?

and Rones (1934)⁵⁸² found that the anterior bulge of the lens was made up of a lenticular-shaped mass of homogenous-staining material behind a normal capsule and epithelium. This material, separat-

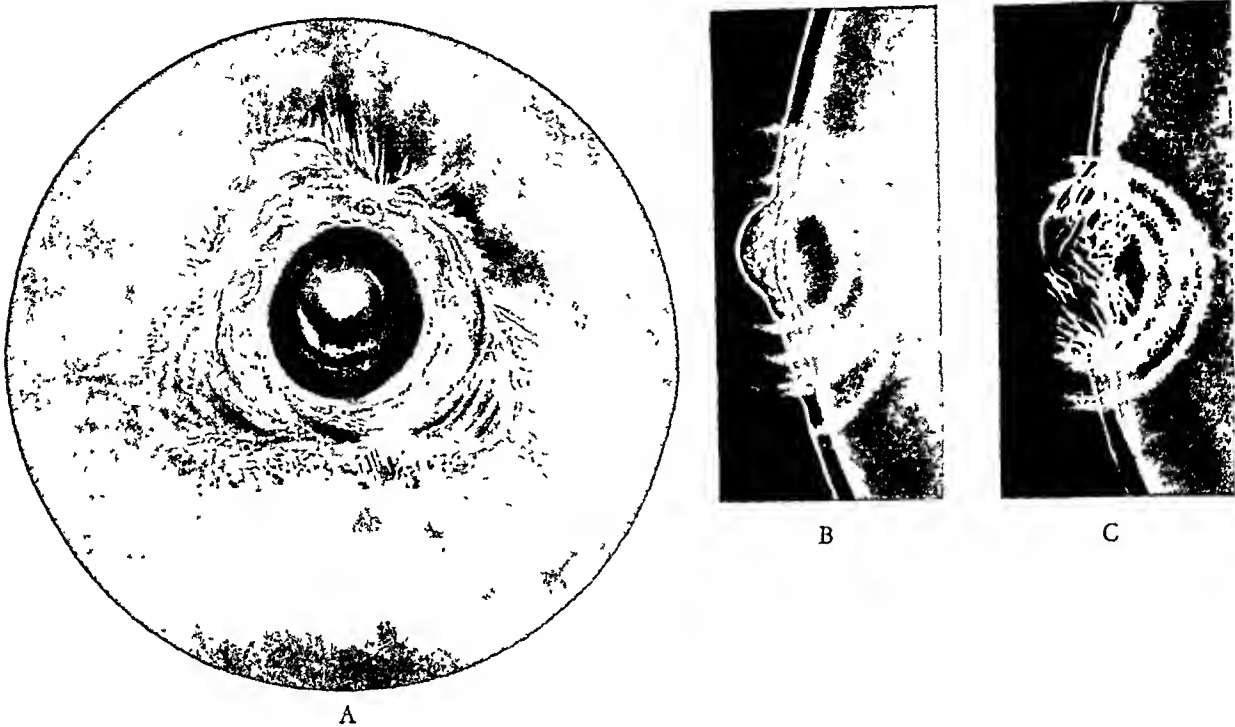


FIG. 376. Anterior lenticonus. A. Diffuse view. B. Before rupture. C. After rupture of the cone. B and C are optic section views. (After Ehrlich.)

ing the latter from the underlying lens fibers, could easily act as deterrent to the ingrowing fibers. Pressure of the neighboring growing fibers could then cause a herniation or ectasia. On the basis of what is seen in certain lower forms (fish and primitive toads) in which the lens appears to bulge through the pupil as though the iris were pressing it back peripherally, Mann has hypothesized that "it is possible that some such deforming stress may have occurred in fetal life owing to a too-rigid pupil, and the lens may have been permanently moulded." Through the courtesy of Dr. Ehrlich, I saw a 10-year-old boy who had a small polar opacity in the left eye (vision 20/30) and who had what appeared to be a progressive anterior lenticonus in the right eye (vision: hand movements). In this completely cataractous eye in the axial region, the base of the anterior cone was outlined by a circular band, which under high power

was seen to be formed by small yellowish white dots (Fig. 376). The interior of the protrusion was composed of fine opaque fibers arranged in irregular layers. Later the top of the globular protrusion

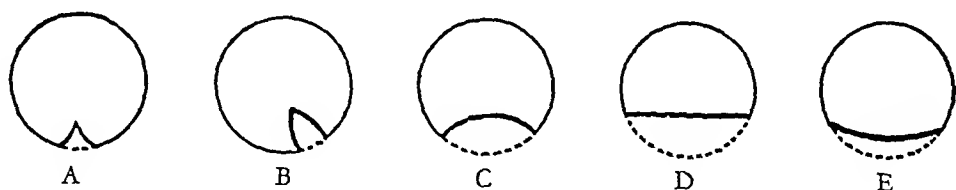


FIG. 377. Colobomas of the lens. Types of defects. A. Notch. B. Triangle. C. Elipse. D. Segment. E. Spindle. (After Kaempfer.)

spontaneously ruptured and some of these fine white fibers (opaque lens fibers) extended from it with their distal ends floating freely in the anterior chamber.

Coloboma of the Lens. This by itself is a very rare anomaly and in most of the reported cases was associated with other defects, especially ectopia lentis and spherophakia. Modern interpretation of its pathogenesis, based on clinical as well as experimental evidence, seems to point to secondary defects or absence of the zonular fibers. In practically all cases observed biomicroscopically the zonule or its remains seen in the colobomatous area was pathologic (fragile, torn, or sprinkled with pigmented or whitish deposits). As previously mentioned the normal flattened axial development of the outer portions of the lens depends in part on the zonular tension. Without this (as seen in spherophakia and ectopia) the lens becomes less flat or spherical. However, it seems in some cases that an added factor of some kind would be necessary to produce an actual coloboma. According to Mann (after Hess and others) persistence of the capsulo-pupillary vessels could cause a zonular defect and that this, in view of the localized absence of zonular traction, would lead secondarily to a colobomatous defect in the lens. Rones suggests that since the growth rate of the lens is not constant, loss of inherent energy in a particular zone may be a factor.

Depending on the extent, colobomas may be small or may involve as much as one-third of the lenticular circumference (Fig. 377).

The smaller ones usually appear as notches while the larger ones may have an elliptical or crescentic shape or a segment defect as if the lens was cut by a chord of the lens circumference.* Coloboma lentis occurs more frequently in the lower part of the lens and when small requires dilatation of the pupil for its detection. In other cases it may be associated with coloboma of the iris and choroid. As in slight subluxations, small peripheral notches may not disturb vision and, unless the pupil is dilated, may be missed. Larger ones may result in aphakic hypermetropia or (owing to increase in spherical shape of the lens) lenticular myopia. The frequent association with opacities (zonular or lamellar cataract, coronaris, etc.) may further affect vision.

DEVELOPMENTAL CATARACTS

CAPSULAR AND POLAR CATARACTS (ANTERIOR AND POSTERIOR)

Capsular cataracts or those just limited to the capsule are rare. With the narrow beam it will be seen that they do not invade the cortex deeply, but because of their thickness they alter the curvature of the lens by protruding slightly toward the anterior chamber (Plate LX, Figs. 3, 4, 5, 6). In contradistinction to polar cataracts they may be found outside the polar regions. The strictly hereditary forms must be differentiated from those acquired pre- or postnatally through inflammation or trauma. Capsular cataracts may be white, but frequently they are sprinkled with small pigment granules which may be found on the surrounding capsular areas as well. As in the case of polar cataracts, capsular opacities may have filaments attached to them. Filaments derived from the pupillary margin would strongly suggest inflammatory synechiae; while those from the lesser circle, a developmental aberration. As a rule capsular opacities are small. According to Koby most of these capsular opacities are pro-

* According to Kaempffer¹⁹¹ the size and shape of the defect depends on the number of the weakened, stretched, or absent zonular fibers. Evidently defects in the more axially inserted fibers (anterior and posterior zonular fibers especially the stronger anterior ones) are liable to cause greater colobomatous defects than the shorter equatorial ones which insert more peripherally. In the latter case depending on the size of the area involved a small notch or larger elliptical defect will result. When both anterior and posterior fibers are entirely absent in an area then retraction of the elastic lens capsule will cause a larger segment-like coloboma extending more axialward.

PLATE LX

FIG. 1. Anterior polar and capsular cataract with reduplications. Pupillary membranes.

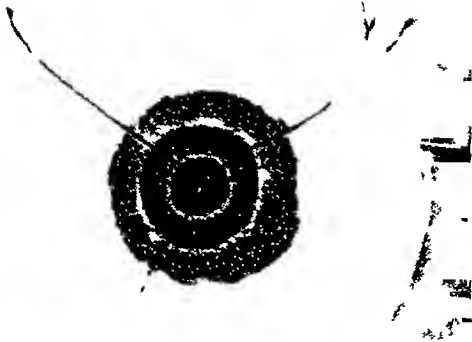
FIG. 2. Same as Figure 1 by direct focal illumination.

FIG. 3. Capsular opacity attached to the iris. Diffuse illumination.

FIG. 4. Same as Figure 3 by direct focal illumination. High power.

FIG. 5. Capsular opacity. Diffuse illumination.

FIG. 6. Capsular opacity, with small imprint. Direct focal illumination.



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duced by old synechiae of iritis occurring before or after birth. Vogt described an unusual case in which a capsular cataract developed after a perforating corneal injury in an eye that previously had well-developed remains of the pupillary membrane.

An interesting phenomenon, greatly stressed by Vogt (1917),⁶⁴⁷ was the presence of a shagreen-free halo surrounding capsular opacities (Vogt's sign). He found this halo in every type of anterior capsular cataract whether congenital, or acquired through trauma or disease. It is not seen with capsular deposits, e.g., remains of the pupillary membrane. Evidently it is caused by a "level" change (characteristic of capsular opacities, or of polar cataracts involving the capsule) in the shagreen field so that specularly reflected rays at this place do not reach the observer's eye. However, by altering the direction of illumination and observation, it is possible to obtain a shagreen in the halo (previously nonreflecting) area. With this maneuver Vogt was able to find microscopic anterior polar cataracts in the form of dots measuring from 0.05 to 1 mm. Two points prove that these little dots were genuine anterior polar cataracts: (1) they were found in individuals whose siblings had anterior polar cataracts or in whom the fellow eye showed one. (2) these dots showed shagreen-free dark halos.

Polar cataracts are situated subcapsularly and may or may not involve the lens capsule. In either case they tend to extend deeper into the lens but not beyond the adult nucleus. In the literature the distinction between capsular cataracts and polar cataracts is not always clear. For example, capsular cataracts that (in the polar region) project into the anterior chamber in the form of a cone or pyramid are known as pyramidal capsular cataracts even in the presence of an imprint in the cortex. I believe that for the sake of clearness the term capsular cataract should be reserved for opacities involving the capsule only and that the complicated ones in which the capsule and the subcapsular areas are affected (in the polar regions) should be known as polar cataracts.

Most of the pyramidal cataracts are of the polar type. Here we see a combination of capsular and subcapsular cataract, in which the

PLATE LXI

FIG. 1. Anterior subcapsular polar cataract (pyramidal). Diffuse illumination.

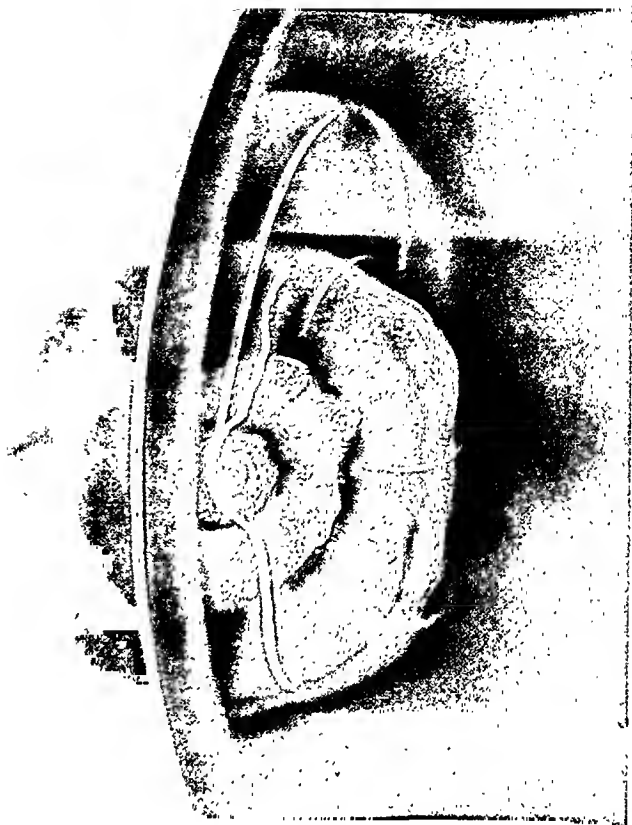
FIG. 2. Same as Figure 1. Direct focal illumination. High power.

FIG. 3. Anterior polar cataract.

FIG. 4. Posterior polar and capsular cataract with unusual imprint seen in the same eye as shown in Figure 3.



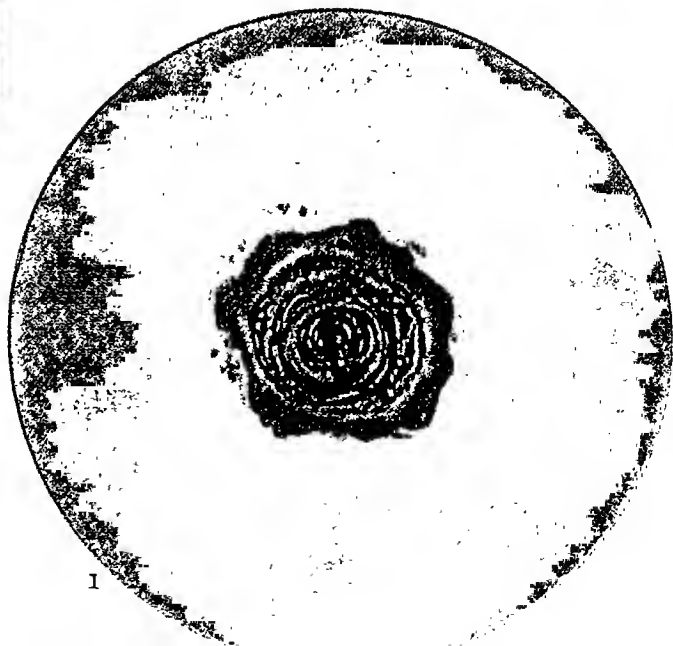
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capsular part may at times be overdeveloped (Plate LX, figs. 1, 2). In some cases depending on the time of origin a pyramidal opacity may develop beneath the capsule, the apex just touching it or the apex of the pyramid may be directed centrally. Whether projecting anteriorly from the capsule or within the lens subcapsularly, these pyramids are generally not solid structures but are formed by terraced layers, starting as a small layer beneath the capsule, each succeeding disk increasing in diameter as the base is approached deeply (Plate LXI, figs. 1, 2, 3). It is easier to explain the formation of these terraces when they occur within the lens (i.e., by the ingrowth of new fibers) than those occurring in the projections that extend pyramidally from the capsular surface into the anterior chamber. One might imagine that owing to a weakness in the capsule, the new fibers beneath the capsule growing in from all sides cause successive layers to herniate because of intralenticular pressure.

Posterior polar cataracts are not to be confused with the hyaloid corpuscle ("physiologic" rest of the hyaloid artery — *cataracta polaris spuria*), which as previously mentioned is not located at the posterior pole but is somewhat nasally decentered from it. Like the anterior polar cataracts with which they sometimes coexist, posterior polar cataracts have varying configuration and size (Plate LXI, fig. 4). Some have dense snow-white centers with fainter irregular borders, while others resemble icicles or are porous or vacuolated, not unlike that of *cataracta complicata*. Their size may also vary greatly from small spots to large disks or saucer-shaped opacities. These disks may be flat, confined to one area, or form pyramids. Reduplication or imprint is common. Just below the capsule they may cause posterior protrusions like a rudimentary posterior lenticonus. As a matter of fact, the latter is frequently marked by polar cataract. Posterior polar cataracts may be complicated by the presence of other types of fetal cataract, e.g., *cataracta centralis pulverulenta*. Vogt reported a posterior polar cataract in a case of bilateral aniridia (aniridia was found in this family in two out of three siblings and was associated with nystagmus and absence of the macula). The polar cataract consisted of two parts, a small subcapsular portion and a

second layer of opacity in the region of the pole of the posterior adult nucleus.

The ingrowth of new lens fibers may separate the opacity partly or completely so as to form an imprint or the so-called reduplication cataract. Partial separation may result in the typical "dumbbell" or "collar stud" shaped opacity, a stalklike process composed of single or multiple strands connecting both ends. The mechanism and time of formation of these opacities is still not exactly understood but many theories including experimental data have been advanced. Depending on its individual characteristics, the period of formation may vary. In certain rare instances the pyramidal shaped opacity may extend forward to be connected with the cornea and according to Mann might indicate an earlier development (at or after the eight mm. stage), the fault lying in an imperfect separation of the primary lens vesicle from the surface ectoderm. However, in most cases (and with more convincing evidence) it is now believed that the development of this anomaly occurs at a later stage. Two chief factors seem to point to this. First the fact that without exception, in cases of polar cataracts the inner fetal nuclei with their Y-sutures are found to be normal. It is hardly conceivable that this part of the lens could remain normal if there was interference with or failure of perfect separation of the lens vesicle from the ectodermal surface.* Added to this is the point that polar cataracts never extend to the fetal nuclei, being limited to the central parts of the cortex and adult nucleus.

Secondly the frequent finding of attached pupillary membranes to anterior polar and capsular cataracts.† This could place the development of these opacities at a later time when the pupillary membrane is formed (i.e., after the separation and completion of the primary lens vesicle, 25 to 30-mm. stage—8 to 9 weeks). The exact role played by the pupillary membrane on the anterior capsule in the genesis of polar cataracts is not known. A suggestive possibility (Vogt) is that there is some interference with the circulation owing

* Vogt found a case in which an anterior polar cataract occurred in association with punctate fetal nucleus opacities. This is perhaps an accidental or coincidental association.

† Both Horner⁴⁸¹ and Terrien⁶³¹ reported cases in which threads were seen attached to posterior polar cataracts. In these cases there were no injuries or signs of inflammation.

to faulty anastomosis or too early degeneration of the vessels of the central areas. However, it is also possible that disturbances in these vessels may result from fetal inflammations. The deeper displacements of the opacities and formation of imprints in polar cataracts can be best explained by the ingrowth of new fibers which in itself is physiologic.*

Some writers have denied the hereditary determination of capsular and polar cataracts. However, without considering those in which the question of frank trauma or secondary disease (as indicated by corneal opacity or signs of previous iritis) it can be said that they are transmitted as dominant Mendelian characteristics. Vogt postulates that according to pedigrees only certain forms (i.e., macroscopic and microscopic, pyramidal or flat-shaped ones, with or without pupillary membranes or imprints) can be regarded genetically as being of the same order, i.e., variations of hereditary polar cataract.† In view of their usually serious damage to vision and dominant transmission these cases represent a problem for the geneticists. However, as is the case with many severe hereditary ocular defects, persons having them are less likely to propagate and hence the defect in itself acts as an eliminating factor.

OPACITIES IN THE VICINITY OF THE FETAL SUTURES

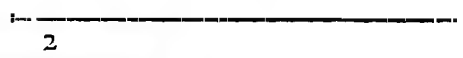
Anterior Axial Embryonal (Fetal) Cataract (Vogt). This form of stationary cataract, although it had been noted previously, was

* As previously discussed on page 1010, the lens is built up of concentric layers composed of fibers deposited one on the other in radial rows (Rabl and Vogt). In this way the apposition of newly formed layers displace, wholly or in part, any opacity located in the anterior parts to a deeper plane. This type of displacement has been observed in other forms of cataract, e.g., zonular, traumatic and in the subcapsular opacities of glaucoma. The zones of discontinuity seem to have a definite relationship to the displacement and also is a preferred locale for cataractous change. In some polar cataracts it may be noticed that the separation or reduplication occurs between these zones and that the densest part of the opacity is located on them (This type of vulnerability is also typical for the sutures). While in others the cataract may cause a fusion of the anterior zones of discontinuity resulting in a convergence of their direction as they merge with the opacity. This probably results from the fact that the opacity (when very dense) prevents the ingrowth of new fibers in these regions, a condition which interferes with the sagittal development of the lens (thickness) as is often seen in special cases and which in extreme cases could result in a form of central umbilication.

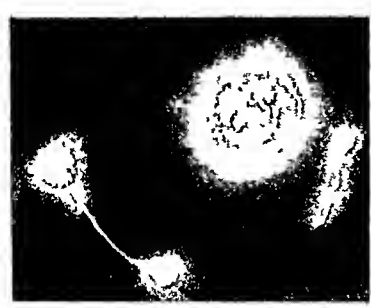
† If the gene for the normal anterior pole is designated as P and that of polar cataract as p then according to Mendel's formula of "back crossing" $PP \times Pp = 2(Pp + pP)$. In other words provided the number of progeny is sufficiently numerous half of them will inherit the defect. In one family of ten children, four were affected, a status where p is dominant to P .

PLATE LXII

- FIG. 1. Form of anterior axial (embryonal) cataract.
- FIG. 2. Form of anterior axial (embryonal) cataract.
- FIG. 3. Form of anterior axial (embryonal) cataract.
- FIG. 4. Anterior suture (stellate) opacities.
- FIG. 5. Posterior suture (stellate) opacities.
- FIG. 6. Unusual type of suture (stellate) cataract. Diffuse illumination.
- FIG. 7. Unusual type of suture (stellate) cataract. Direct focal illumination.



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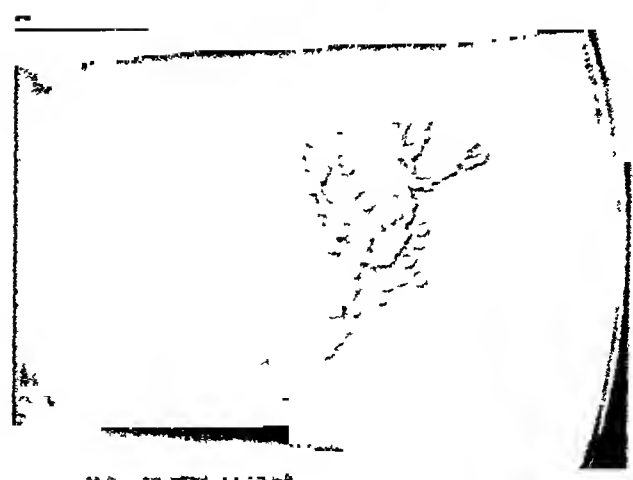
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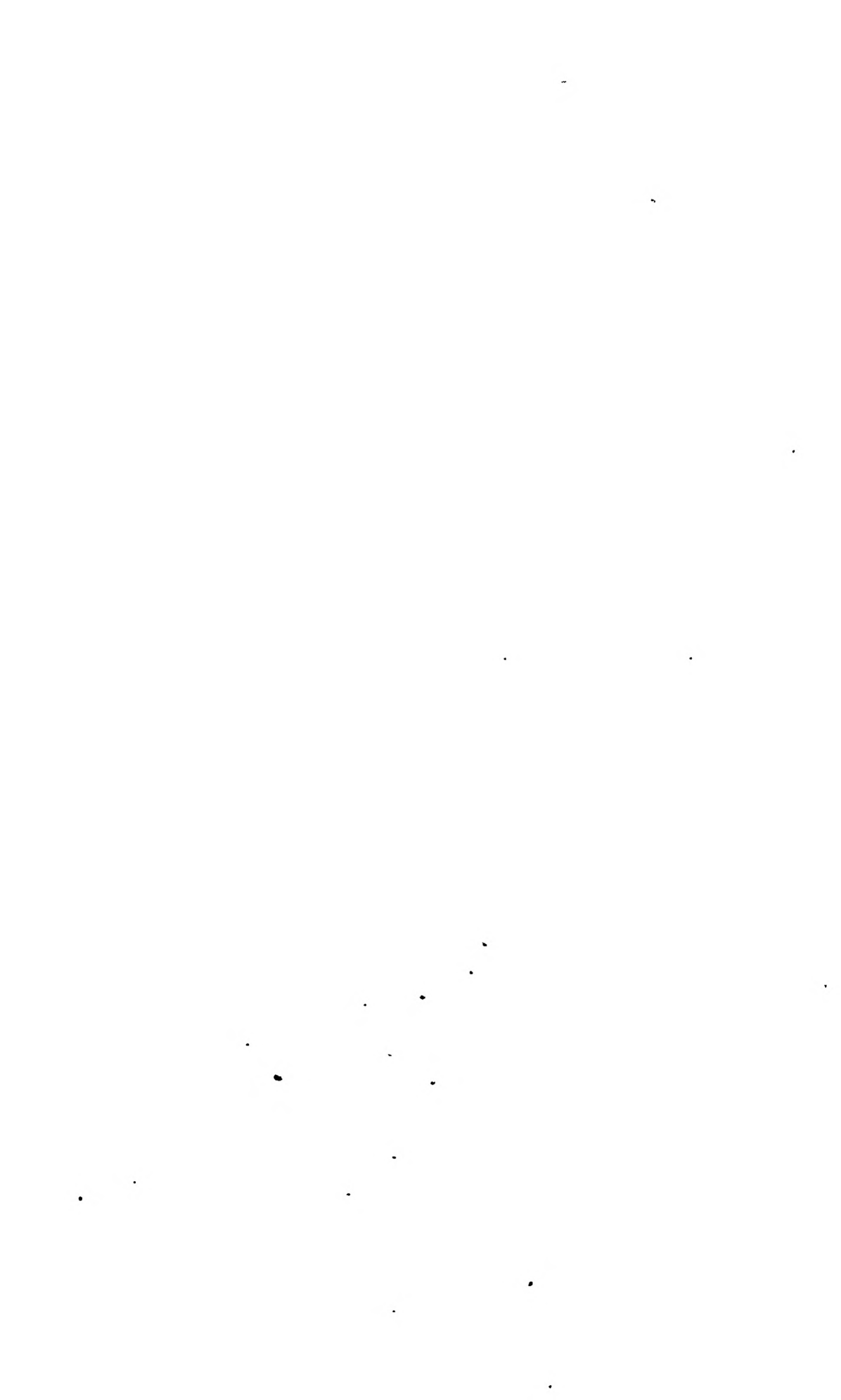


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not completely understood until 1918 when Vogt, employing the narrow beam, described it as an entity. Because of its axial location, it can be easily seen even with the undilated pupil by means of the optic section, narrow angle, and careful focusing of the microscope in the central area using the dark interval as a guide. It occurs in from 20 to 25 per cent of normal persons but is not necessarily bilateral. The whole figure and the component parts are so slight that it does not ordinarily interfere with normal vision. Its appearance and location is so distinctive that errors in diagnosis are hardly likely. Characterized by great delicacy, it consists of fine intensely white dots, occurring singly or in groups, invariably located within or in the immediate vicinity of the anterior Y-suture (Fig. 378; Plate LXII, figs. 1, 2, 3). Only rarely are they located within the central dark interval. With age they may appear slightly yellowish in color. The design of the opacity can be formed by isolated or contiguous small groups of dots, which may be joined to one another by fainter nebulous veils. The opacities may or may not lie in one plane, usually in that of the suture, or if not some of the individual groups may be found slightly anteriorly or posteriorly, the intervening distance generally being minute. The little opaque areas are formed of the finest white dots held together by a delicate veil-like opacity some times no more marked than if they were a misty halo. Occasionally the groups situated at the ends of the arms of the Y will serve to accentuate the suture or, if more extensively developed, may outline it completely (Fig. 378). The opacities veiled by a surrounding haziness, which outlines the ends of the suture arms, may appear rounded and may glitter like a hanging jewelled pendant supported by a delicate chain (the suture) (Fig. 378). At times, this type of cataract may only be indicated by a few isolated dots, irregularly distributed or in the form of short lines or in a circular disposition (Fig. 378). Because of its central location, the time of its formation was thought to be in the early period of the formation of the primary lens vesicle. Actually it is found to be connected with the anterior Y and not with the central interval. This suture first becomes apparent at the beginning of the third month. During the third month the anterior

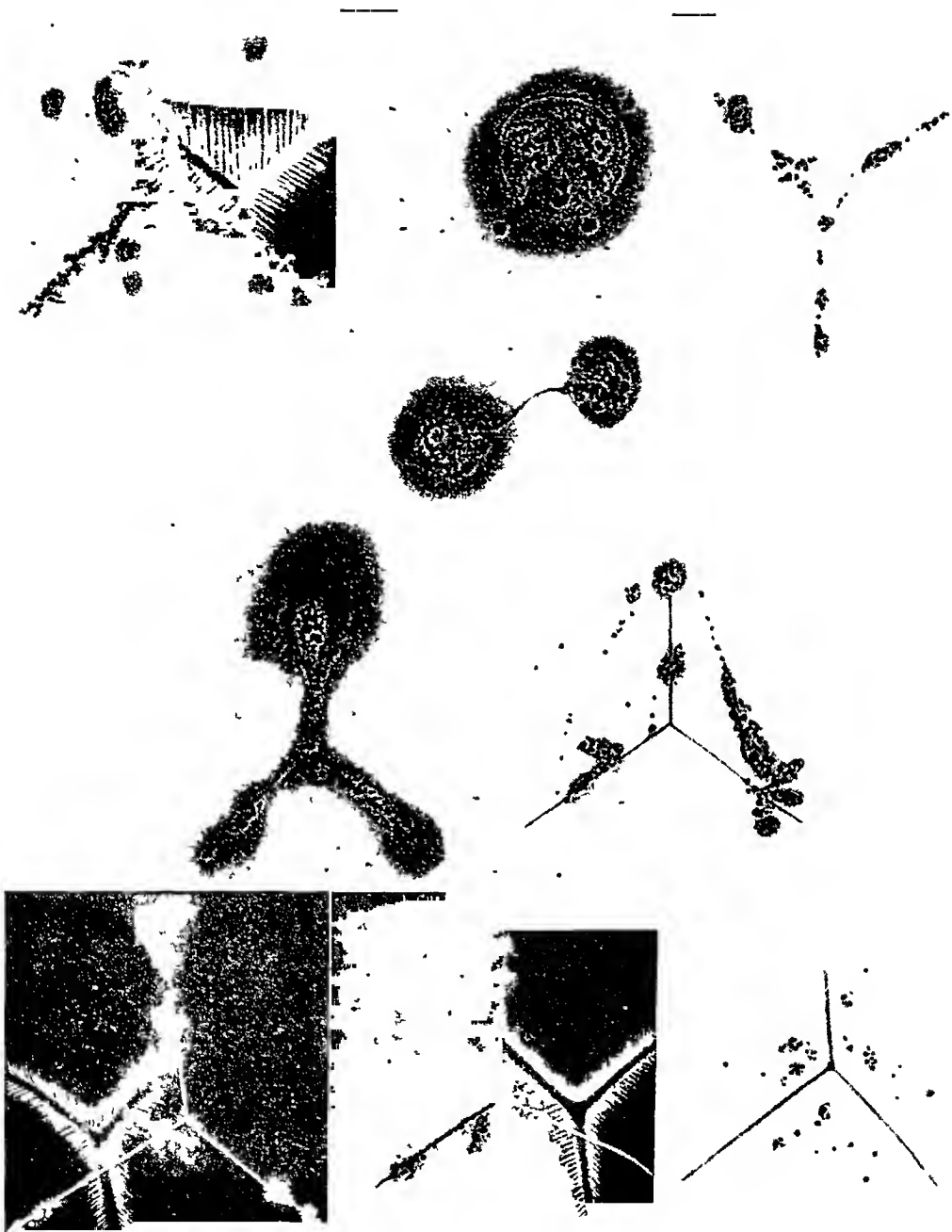


FIG. 378. Types of anterior axial embryonal (fetal) cataract (After Vogt)

Y is completed, but the lens diameter at this time is only about 1 mm. Since the axial embryonal cataract can occupy a region measuring from 1 mm. to 2.5 mm., its earliest development (according to Vogt) should be traced to the late part of the third month or early part of the fourth month. Because of its frequency, even in otherwise normal lenses, this type of opacity is commonly found accidentally associated with other types of congenital and acquired cataract. Vogt observed that in some pedigrees the anterior axial embryonic cataract is a hereditary dominant and suggests that the frequency of this hereditary type can be explained by the fact that opacities that do not affect vision do not have an eliminating value.

The hereditary character of this opacity was further exemplified in the study of identical twins. In two sets — eight eyes — the type of opacity was identical, differing only slightly in number and distribution. In these identical twins Vogt also noticed the similarity in the rests on the posterior surface of the lens, i.e., the arcuate line and floating hyaloid remains. In his opinion the reabsorbing processes in the region of the posterior pole of the lens (in normal eyes) are governed by heredity. This would add another "idio-variation" to those known to depend on the "normal" mechanisms of hereditary processes, e.g., curvature of the cornea, length of axis of the eyeball, pigmentation of the iris, development of the iris frill, shape of the pupillary pigment seam, shape of the disk, and course of the retinal vessels, etc.

Stellate Cataract (Anterior and Posterior) (Synonyms: Cataracta stellata, triradiate or sutural cataract.) Stellate cataract is a rather striking axial opacity invading the anterior or posterior fetal sutures, either singly or together. It delineates the suture distinctly and when marked may be observed at times even with the unaided eye. Cataracts of this type tend to have a solid appearance, their thickness and width varying from place to place irregularly, becoming especially wide at the places of branching. In some cases the edges of the suture opacity are scalloped, giving it a festooned appearance. The branches or arms of the suture cataract may measure up to 1.5 mm. in length making the total size of the figure relatively large.

Frequently each individual suture cataract may be composed of a double layer, each layer differing in width, thickness and particularly in color. In some cases these layers appear as if they were fused one

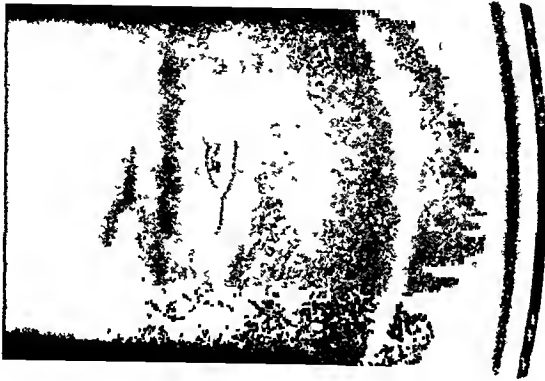


FIG 379. Stellate (anterior) cataract. (Resembles anterior axial embryonal cataract.)

on the other but in others it will be seen that one arm or another lies a little in front of the corresponding deeper one. In any case they do not form an exact replica of each other. Often, small dots or disklike opacities will be found in the vicinity of these figures. The deeper figure of each suture cataract (anterior and posterior), i.e., the one closest to or directed toward the dark interval, is more transparent and hence appears bluish or greenish in color (a phenomenon not unlike that seen in cerulean cataracts) (Plate LXV, figs. 4, 5, 6, 7). The intensity of this color display depends upon diffraction and varies with the change in angle between illumination and observation. The blue or anterior part of the posterior suture cataract corresponds in outline to the Λ and lies in front of it. On the other hand the deeper part of the posterior suture cataract is densely white and is usually wider and longer in extent. The location of the smaller, bluer figure anterior to it corresponds to an older segment and by the time of formation suggests a separate development (Vogt). Not all stellate cataracts have this double formation. There are instances when the opacity is all blue or greenish, or others where it is completely white. Likewise, its consistency may vary,

being mosslike or feathery, approaching that of cataracta dilacerata (page 1101). It is not unusual for the stellate type of cataract to co-exist with other forms of developmental cataract, e.g., zonular, cerulean, and dilacerata, although it may be found in an otherwise normal lens.

When a stellate cataract is confined to the anterior Y-suture only and is poorly developed, it might be confused with anterior axial embryonal cataract (Fig. 379). Differentiating features are (1) the delicacy of the anterior axial embryonal cataract, which is composed of small groups of dotlike opacities, frequently surrounded or enclosed by the characteristic veil, and (2) the fact that it generally is located closer to the dark interval. The not infrequent concomitant presence of a posterior stellate cataract would put one on his guard, although it is conceivable that an anterior axial embryonal cataract might be found coincidentally with a posterior stellate cataract. The genesis and time of development of the stellate (suture) cataract, which is essentially stationary, is not known; owing to its location and size it is generally believed to form late in fetal life, and evidently later than the anterior axial embryonal cataract. Vogt believes that deflections or branching of the ends of the arms of the suture cataract (corresponding to the normal development of the sutures in man as against the lack of branching in lower animals) represents a sign of later development of the cataract. The branching of the suture ends normally occurs at a later period in the development of the sutures.

CONGENITAL DIFFUSE NUCLEAR CATARACT (VOGT)

Vogt (1931) described what he considered to be a new picture of a hereditary condition affecting in a total way the fetal nuclei. Unlike zonular cataract, which is made up of punctate opacities and hooklets, this opacity consists of a diffuse structureless haze not unlike that of the senile nuclear cataract (Fig. 380). 'It differs from the latter, however, in that by outlining the shape of the fetal nuclei (inner and outer) its form is more curved and also its color is different. In the past I have seen such opacities in young adults but

considered them to be a presenile or early incipient nuclear cataract rather than congenital. But since all the so-called presenile and senile changes may be genetically determined, it is not surprising that this type of cataract should appear congenitally.

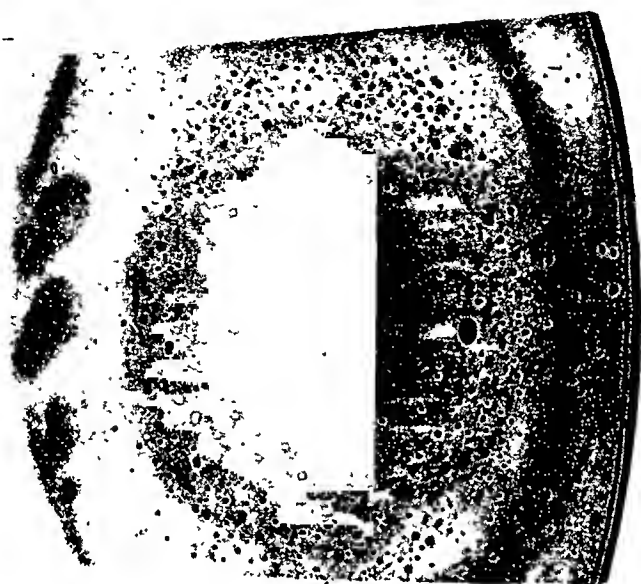


FIG. 380. Congenital diffuse nuclear cataract combined with cerulean opacities.

Vogt described cases of this type of cataract in a 4½-year-old girl and in her mother, aged 34 years. In the girl, optic section showed a large round opaque yellow-green nucleus. Peripherally the zone of discontinuity of the cortex was markedly divergent. With the ophthalmoscope, a rounded shadow dislocated slightly upward was seen in the pupil. The vision was 3/20 in each eye. The mother had a similar finding, except for a pointing of the nucleus in the region of the equator, a normal finding in the adult. According to Vogt this may be an effect of zonular tension. Also corresponding to her age, the cortical thickness was greater in the mother, and because of shrinkage, the nucleus was smaller. There was a fainter layer of more grayish or bluish relucency in the peripheral cortex. In both there were anterior axial embryonal opacities. With the ophthalmoscope the central shadows in the mother were somewhat denser than that in the child. Vogt found similar opacities in the mother's sister

and brother and consequently considered it as a dominant hereditary disease of the lens nucleus.

CONGENITAL, EMBRYONAL OR FETAL NUCLEAR CATARACT
(CATARACTA CENTRALIS PULVERULENTA AND DISCIFORM CATARACT)*

This form of congenital nonprogressive cataract which is composed of small punctate dots of varying sizes arranged in the shape

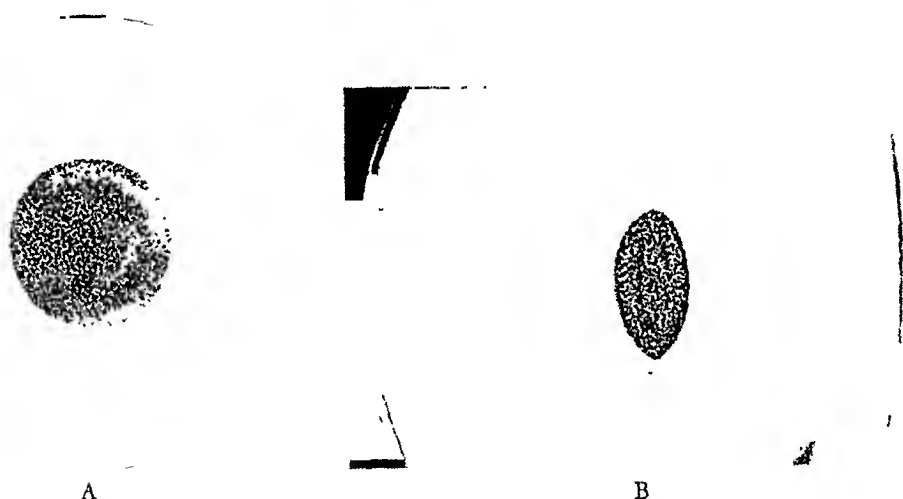


FIG. 381. A. Cataracta pulverulenta (congenital fetal nuclear cataract). Diffuse illumination.
B. View of above by means of the optic section. Direct focal illumination.

of a disk, lies in the central part of the lens, i.e., in the fetal nucleus. It has been suggested that it is a *formé-fruste* of the larger zonular cataract, especially since alternating occurrence of the two types have been seen in the same family. It is practically always bilateral but is rarely of sufficient density to greatly interfere with vision. Apparently this type of central cataract was inaccurately localized by many writers in the posterior parts of the lens before the days of biomicroscopy although they realized its hereditary (ideokinetic) and nonprogressive nature. Nettleship and Ogilvie † (1906) published an extensive report on this type of cataract, occurring in the descendants of a certain John Coppock born in 1774 in Oxfordshire, England. Earlier, Doyne reported several cases in this same family.

* Also possibly hereditary forms of the smaller central zonular (lamellar) cataract; disciform or Coppock cataract (Nettleship and Oliver), Doyne's discoid cataract, family nuclear cataract.

† Nettleship, E., and Ogilvie, M. F. "A Peculiar Form of Hereditary Congenital Cataract," *Tr. Ophth. Soc. U. Kingdom* 26:191 (1906).

PLATE LXIII

FIG. 1. Central discoid fetal cataract. Right eye. By diffuse illumination.

FIG. 2. Central discoid fetal cataract. Same as Figure 1. By direct focal illumination. Unusual form.

FIG. 3. Partial form of the central discoid fetal cataract. Left eye. By diffuse illumination.

FIG. 4. Same as Figure 3. Direct focal illumination.

FIG. 5. Cataracta centralis pulverulenta associated with anterior lamella opacities. Diffuse illumination.

FIG. 6. Same as Figure 5. Direct focal illumination.

FIG. 7. Central discoid similar to case in Figure 1.

FIG. 8. Congenital central punctate opacities. Similar to case shown in Figure 5.



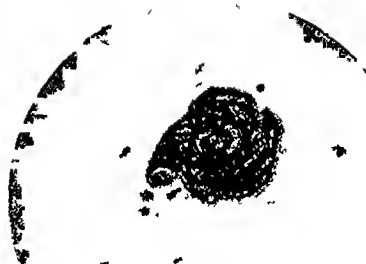
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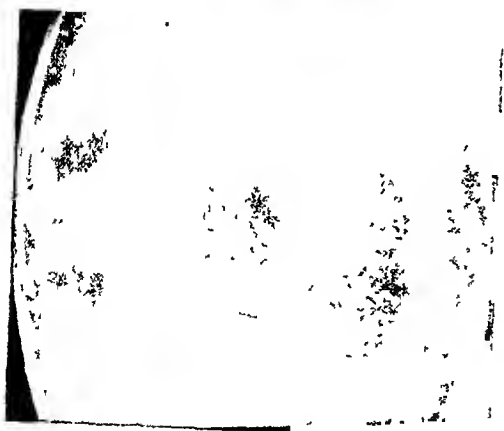
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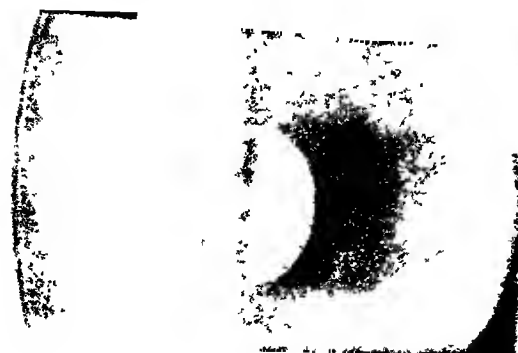
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Other cases, not related to the Coppock family, were mentioned by Parsons and Harman. Vogt,* in 1921, unaware of the English reports, described a similar form as a new variety of congenital cataract, which he termed *cataracta centralis pulverulenta*. Later, Gifford† (1927) pointed out that *cataracta centralis pulverulenta* was probably identical with Coppock and Doyne's cataract.

Ordinarily, it is shaped like an ellipse and is made up of fine points (Plate LXIII, fig. 6). These points vary slightly in size and are usually most condensed in the central part of the opacity where they may obscure the Y sutures. In others a concentric ring form may be seen in which the central part is composed of more irregular, larger spots, making them whiter in appearance. This is surrounded by one or more rings of finer dots. Vogt has illustrated several variations of this lesion; among them is a case in which white points were found in front of the central interval, or the place where the anterior axial embryonal cataract is localized. There were no opacities in or behind the dark interval. Some of the spots were surrounded by a small halo. In the periphery of the area occupied by them they were arranged in lines forming a figure suggestive of an irregular star. In another case, there was a lens-shaped opacity, the equatorial diameter of which measured 0.5 mm., located with the narrow beam a little in front of the center of the dark interval, filling the anterior region of the dark interval and that of the anterior Y-suture, which could not be seen. The posterior A behind was visible and uninvolved. In the periphery the dots, in contrast to the above-described case, were not linear but gradually faded into the surrounding unaffected areas. There were no concentric layers but the equator of the opacity was rounded or circular. By strong magnification the dots appeared as brilliant white platelets of varying size. The largest were 0.05 mm. and the smallest were like dust. The thickness of the entire opacity was estimated at about a little more than half the equatorial diameter. In the zone of discontinuity corresponding to the adult nucleus a second but less conspicuous zone of opacity was seen. About

* Vogt, A., *Hand. der Spaltlampe Microscopie*, 1921.

† Gifford, S., "Zum Kongenitalen Star des Embryonalkerns," *Klin. Monatsbl. f. Augenb.* 28: 191 (1927).

15 hooklike opacities or riders were distributed on the circumference of the nucleus. Bellows³⁶⁴ also reported such a finding in a 36-year-old man with normal vision, in whom a central area, measuring 2.5 mm. was surrounded by an outer ring of hooklike radial opacities measuring 4.5 mm. in diameter. Because of this not too infrequent combination, i.e., cataracta centralis pulverulenta and hooklets of the adult nuclear zone, it is possible that this type of cataract may represent a transitional or earlier (hereditary) form of zonular cataract. In a third form he noted a ring of fine dots forming a concentric layer around the anterior Y-suture behind which was a small delicate opacity (disklike) that glistened somewhat and measured about 1 mm. in vertical diameter. The outer ring measured 2.7 mm. These variations are all similar to the typical opacity of cataracta centralis pulverulenta in that they involve the central interval, they measure no more than 1 to 3 mm. in diameter, and they are all composed of fine dots and dustlike opacities, which are not dense enough to interfere with normal visual acuity. I recently have observed an unusual type in which the anterior Y itself was heavily infiltrated not unlike the stellate cataract. Vogt reasoned that since the lens vesicle has an equatorial diameter of about 0.5 mm. in the third month and from 0.9 to 1.4 mm. in the fourth month, the opaque part of the lens ordinarily would correspond to the size of the latter at the third month. This corresponds to the time of beginning development of the pupillary membrane and of a consequent change in the manner of the lens nutrition. An interference with lens nourishment might conceivably result in this type of cataract.

Vogt found forms of cataracta centralis pulverulenta three times in two generations, indicating a dominant transmission. In Russo's cases⁵⁸⁷ two brothers were affected. Following a consanguinous marriage of one of the brothers, the three sons had these cataracts. The children of the nonconsanguinous marriage of the second brother were free of this defect. According to this, the pulverulent opacities in these cases would be of a recessive type. However, A. Rados (Archives of Ophthalmology, July 1947, Vol. 3 P, pp. 57-77) in an excellent study of central pulverulent cataract and its hereditary

transmission, questions the validity of Russo's assertion that in his family there was an apparently Mendelian recessive inheritance. From a detailed study of pedigrees Rados showed fairly conclusively that "In the hereditary form of central pulverulent cataract, the dominant mode of inheritance is to be expected in accordance with the knowledge of inheritance of various forms of hereditary cataract."

On several occasions I have seen another discoid type located in the same area of lens which instead of being composed of dots was characterized by more solid and opaque lamellar-like leaves (Plate LXIII, figs. 1, 2, 3) surrounding the fetal nucleus. The disk may be complete or only partially formed (Plate LXIII).

ZONULAR (LAMELLAR) CATARACT

The zonular, or lamellar, cataract in its common form appears as a central discoid opacity. Ordinarily it occupies the outer fetal nucleus or areas just outside it and consists of fine white points and peripheral riders. One might compare it to the kernel of a nut, the outer shell representing the adult nuclear stripe. There are many exceptions both in regard to composition and location. These will be dealt with later. As compared to other forms of cataract in the young, its frequency is high. In the past some writers went so far as to state that it is the most common type of lens opacity in youth. This is understandable since in most cases zonular opacities are comparatively large; they occupy a greater portion of the central area of the lens and, consequently, affect vision seriously. In addition they are easily recognizable (Fig. 382). With the dilated pupil, they can be diagnosed macroscopically. But since the advent of biomicroscopy, the focal beam has revealed a higher incidence of the more delicate congenital opacities, such as anterior axial embryonal cataracts and cataracta centralis pulverulenta. These do not affect vision and unless they are unusually dense, they cannot be seen with the ophthalmoscope or by diffuse illumination.

Zonular cataracts may be congenital or acquired. They are generally bilateral; only about 5 per cent of cases are uniocular. They may

arise pre- or postnatally. The congenital ones are either hereditary* or are caused by exogenous influences which may correspond to those known to cause zonular cataract after birth (e.g., hypocal-



FIG. 382. Zonular cataract.

cemia [tetany cataract]). Congenital zonular cataracts may also be found secondary to fetal inflammation or in association with other malformations, e.g., microphthalmos, pupillary rests, iris colobomas, etc. Probably, a considerable number of the zonular cataracts arising prenatally in the first few years of life are associated with infantile tetany (see chapter on endocrine cataracts). A similar relationship has also been found in cases of zonular cataract occurring in older individuals (Meesmann). Apparently they may arise in cases of spontaneous latent tetany as well as in those following inadvertent damage or removal of the parathyroids in operative procedures on the thyroid (see endocrine cataracts, page 1185). In addition, especially in the young, a type of zonular cataract may develop after trauma (page 1257).

The optic section permits localization of the involved areas and visualization of the internal structure (inner fetal nucleus and dark

* Perhaps even these are genetically determined only in the indirect sense that they follow a hereditary defect in other organs (hormonal) which in turn could result in toxins or interference with lens metabolism.

interval may or may not be involved—Fig. 383); many of the variable details found in this type of opacity may also be recognized. As already mentioned the common variety is localized principally

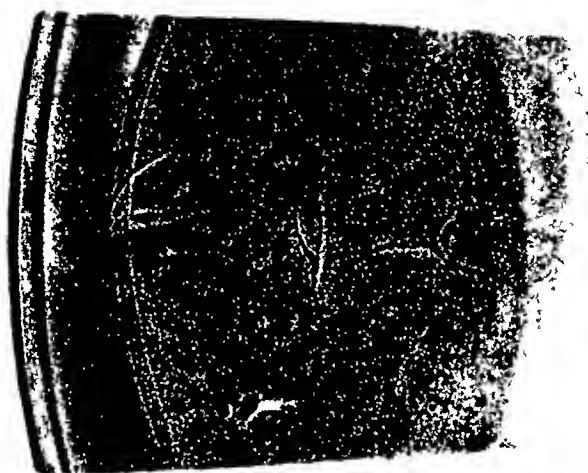


FIG. 383. Zonular cataract.

in the outer fetal nuclei and the deeper layers of the so-called "adult nucleus" and is usually composed of fine white dots, although at times larger flattened spots may be seen. A thin veil-like envelope may surround the main central opacity. In front view, this envelope may be represented by one or two thin concentric lines separated from the central opaque disk by a narrow clear area (Plate LXIV). At times the envelope is thicker and may be made up of fine white points. Extending at right angles to the envelope and often contained in it are the characteristic "hooklets" or riders. In only rare cases are they absent. Sometimes larger riders extending some distance from the equator alternate with smaller ones that are closer to it. Isolated ones are found around the equator of the opacity but are also separated from it. The location of the riders frequently corresponds to that of the suture system but may in addition be situated in between them. These riders often have pointed ends and are very white in color. They may be formed from individual fibers or bundles of fibers that have become isolated from the main opacity. Vogt has described several different pictures. He has reported that

PLATE LXIV

FIG. 1. Zonular cataract. Illustrating central punctate opacities. Surrounded by a delicate envelope and hooklets. Diffuse illumination.

FIG. 2. Same as Figure 1. Direct focal illumination. Optic section.

FIG. 3. Zonular cataract. More solid type.

FIG. 4. Variety of lamella (zonular cataract). Right eye. Diffuse illumination.

FIG. 5. Optic section of case in Figure 4.

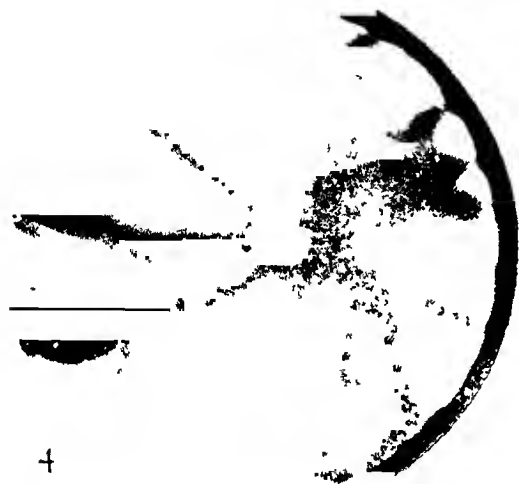
FIG. 6. Left eye of case shown in Figure 4. Diffuse illumination.

FIG. 7. Linear type (incomplete) of zonular cataract. Diffuse illumination.
(Courtesy of Dr. Webster.)

FIG. 8. Direct focal (optic section) view of case shown in Figure 7.



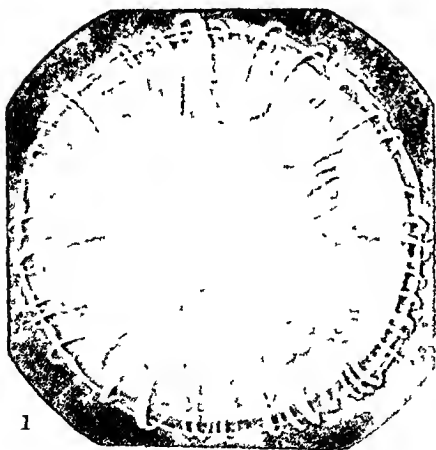
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the sagittal thickness of lenses having zonular cataracts is less than normal, indicating lag in development in lenses having this type of cataract. In some cases radial slits or clefts are seen in the peripheral

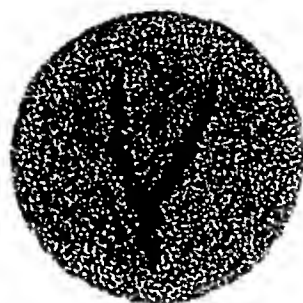


FIG. 384. Cataracta pulverulenta with stellate or suture cataract of the anterior Y suture. Note its small size as compared to zonular cataract.

parts of the opacity, some of which extend to the equator of the opacity where they appear like wedge-shaped defects. The sutures themselves may not be visible. The bases of the wedge are directed toward the periphery (the equator). A radially directed rider may occur in the clear part of the wedge-shaped crevice, its curved part extending out from the crevices in the region of the envelope. At times these crevices may be closer to the axial parts of the lens, but even these will be found to contain a linear streaklike opacity (rider) whose direction corresponds to that of the wedge. In one case of this kind polar cataracts occurred which connected the capsule to the central part of the opaque nucleus. In another case a dense central zone, made up of dots, was surrounded by one of lesser density occupying the adult nuclear zone (envelope), composed of concentric and radiating opaque lines and larger flat opacities. Within the inner disk was an irregular layer of crystals. The inner disk measured 5.2 mm. and the total diameter of the outer one was 8 mm. The great size of such a lens opacity would indicate postnatal development, since it is much larger than the normal fetal nucleus. In some in-

stances zonular cataracts are marked by the presence of a well-developed suture system. This form distinguishes itself by a sharp equatorial margin (without an envelope), by punctate opacities with a clear center (inner fetal nucleus), and by densely opaque white suture lines at the surface of the opacity. This is the "suture" type (*suturata*) to be distinguished from the aforementioned "cleft" type.

The inner fetal nucleus may also be involved, presenting a central punctate opacity within the outer one. They may or may not be separated from one another by a clear interval. The central dark interval (embryonal nucleus) may be free of opacities, leaving a dark slitlike area between the affected coffee bean-like nuclei. In other instances, the dark interval contains punctate opacities, and the inner fetal nucleus is clear. When this is the case, there will be a greater distance (dark) between it and the outer affected nuclei. Another interesting variety is that in which the surface shows neither sutures nor punctate opacities but is made up of an irregular concealing layer of opacities containing radiating lanceolate lens fiber clefts. Vogt states that this type resembles the subcapsular layer forms seen in tetany cataract.

The combination of zonular cataract and presenile (*coronaria*) and senile cataract is not unexpected. The combination of polar cataract and zonular cataract seems to justify the opinion that certain zonular opacities are hereditary. Vogt writes that he has seen a case of total cataract in early youth develop into a zonular opacity through the apposition of new fibers. In addition to the well-developed zonular cataracts, there are certain forms of opacities which, because of their location (outer fetal nucleus) and lamellar character, are to be considered as abortive (*formes fruste*)—or at least related to—zonular cataracts (e.g., congenital nuclear cataract). These may be seen as small, localized layer opacities only partially outlining the outer fetal nucleus (Plate LXIV). Some are axially located and others, outline the equator of the fetal nucleus. An especially interesting case of this type in an exaggerated form was recently called to my attention. It occurred in a 34-year-old man

whose sister had tetany cataract (referred to on page 1186). In both of his eyes there were flattened leaflike opacities located just outside the fetal nucleus. The flattened leaflike structure extended around the equatorial border of the nucleus (similar to zonular cataract), the anterior ends curling like the petals of a flower in the process of opening. The opacity itself was white, shiny, and lardaceous, showing no punctate structures.

OTHER RARER FORMS OF CONGENITAL AXIAL CATARACTS

Certain of the rarer axial congenital cataracts were described before the discovery of the biomicroscope (e.g., coralliform, and axial fusiform cataracts), while others like the spear cataract, cataracta pisciformis, and the floriform cataract were described and named afterward. In prebiomicroscopic days there was no possibility of exact localization, as we now understand it. The suture system and zones of discontinuity (as seen in optic section) and their significance as regards the normal embryological and postnatal development of the lens were unknown. Consequently, using more or less diffuse illumination or transillumination (ophthalmoscopically) only surface or frontal views of lens changes were obtained. This naturally resulted in a morphologic classification based on frontal appearance alone. With the advent of the biomicroscope, two added features in diagnosis became possible: (1) the ability to see through opacities and other changes sagittally and (2) exact localization of the lesions or, better, correlation of their position with the zones of discontinuity or growth rings of the lens. Today we are better able to approximate the time of formation of defects in relation to the ontogeny of the lens. There is still much confusion in the literature concerning these rare forms of congenital axial cataract as is seen by the overlapping descriptions and by the different names that have been given to apparent variations of the same forms. These opacities may or may not be confined to any single zone or zones of discontinuity. Some are localized in the outer fetal nucleus or in the adult nucleus; some extend outward from the fetal nucleus and pass into the adult nucleus or even into the cortex to the

neighborhood of the capsule (coralliform, fusiform, and pisciformis, etc.). Although these opacities differ one from the other in structure, extent and location they have two important common characteristics: (1) they are of a hereditary nature and (2) they are predominantly axially located. Like so many developmental congenital cataracts, no adequate explanation for the mechanism of formation of these types of axial cataracts has been found, especially for those which do not follow the orderly arrangements of the lens structure. Following Knies (1877)⁴⁹⁸ and later Hess (1893)⁴⁷¹ and Collins (1908),³⁸⁶ Mann suggested that abnormal adhesion of the embryonic nucleus (primary lens fibers) occurs to the capsule at the anterior pole * or to the posterior pole, or (as in the case of fusiform axial cataract) to both. Hence, as the lens continues to develop these centrally attached fibers will stretch, become opaque, and prevent the secondary fibers (growing from the equator) from separating the capsule at this point. This they must do in order to meet and to form a normal suture. Failure of separation of the primary fibers from the anterior capsule might result in a coralliform cataract, while failure of separation from both anterior and posterior capsule would lead to an axial fusiform cataract. One might further hypothecate that some of the other rarer forms of axial cataract (spear cataract) might result from modifications of this type of defect, depending on the time of growth of the epithelial cells genetically affected. It is difficult to explain the pathogenesis of the opacities situated outside of the fetal nucleus in an otherwise normal lens except by virtue of their proximity to the capsule in fetal life.

The rarer forms of congenital axial cataracts to be discussed in this section are:

1. Cataracta pisciformis and associated types (Vogt)
2. Coralliform cataract

* The observations of Hess (1893) confirm this idea. He described a chick embryo in which there was a failure in separation of the lens vesicle from the surface ectoderm as a result of which the primary lens fibers grew through an opening in the cornea. Had this closed and had growth been resumed, one might surmise that an axial cataract would have formed.

3. Axial fusiform cataract
4. Spear cataract
5. Floriform cataract

Cataracta Pisciformis. Under the caption "hereditary fish- to fin-like juvenile band cataract" (*cataracta pisciformis*) Vogt described several types of predominantly axial cataracts (Plate LXV figs. 1-6). First, the *pisciformis*, which are axially located, thin, white spotted opacities limited to either the posterior adult nuclear zone or in the juvenile form to the region of the posterior outer fetal nucleus. The ends of the opacity are somewhat curved, giving them the shape of a fin (Vogt). In one case they were associated with a few irregular dots on the adult nucleus and in the cortex anteriorly and posteriorly.

Second, a rare ring of hooklike or zigzag opacities localized to the periphery of the anterior outer embryonal (fetal) nucleus plane. These were found bilaterally in one patient, a young man of 28 years. The zigzag line of opacity was only seen frontally. In section the opacity did not involve the equatorial curvature of the nucleus (as do the hooks in zonular cataracts) but lay like a line on one plane.

Third, ringlike flattened opacities, having a dark central hole (Rosskümmelähnliche) and arranged in radial groups. They are found grouped in the axial region of the adult nucleus and extend peripherally in a radial direction diminishing in size; this radial formation suggests a relationship to the suture system. As Vogt has intimated the appearance of both *cataracta pisciformis* and Koby's later described floriform cataract are similar.

Fourth, spotted cataracts (white or blue) of the anterior and posterior outer embryonal (fetal) nuclear zone. These are groups of axially located small flat spots whose color varies from white to blue, depending on their thickness. Some are irregularly distributed while others assume a radiating form. In a few of these cases there were peripheral cerulean opacities.

Fifth, a blue-green anterior rosette cataract with a layer of white spotted opacities in the anterior and posterior fetal nucleus. In the

PLATE LXV

FIG. 1. Cataracta pisciformis (floriformis). Right eye. Diffuse illumination.

FIG. 2. Left eye of case shown in Figure 1. Note anterior suture cataract. Diffuse illumination.

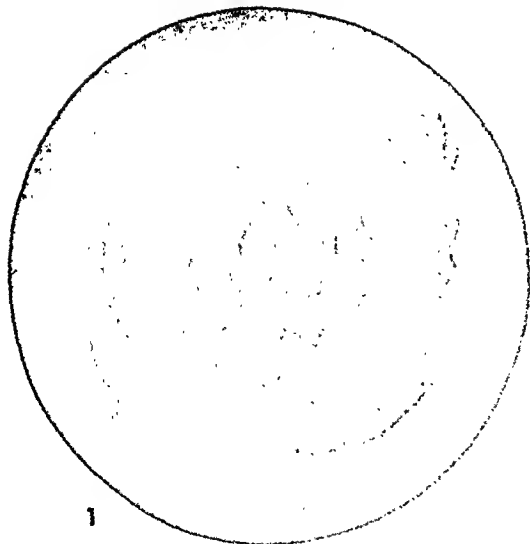
FIG. 3. High-power view of the opacities seen in Figure 2. Diffuse illumination.

FIG. 4. Direct focal illumination of the anterior part of case as shown in Figure 1.

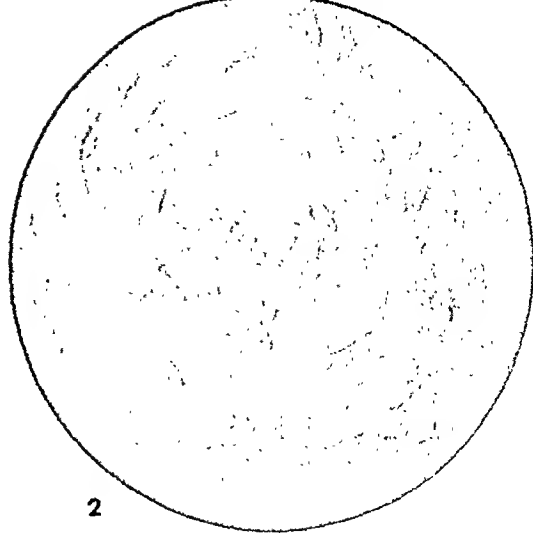
FIG. 5. The opacities in the deeper (posterior part) of case shown in Figure 2. Diffuse illumination.

FIG. 6. Same as case in Figure 5. Direct focal illumination.

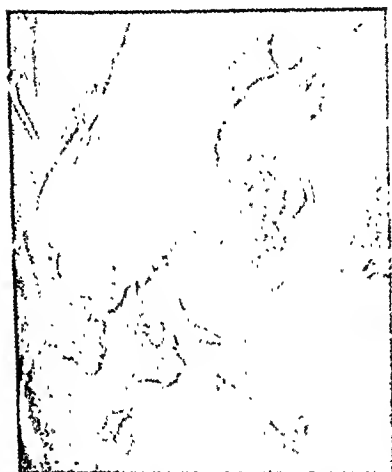
FIG. 7. Unusual form of cataracta pisciformis (floriformis). Diffuse illumination.



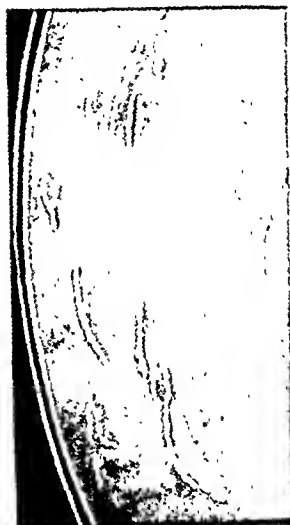
1



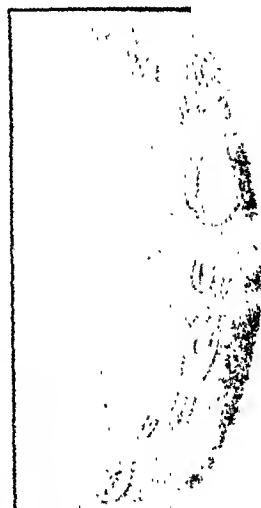
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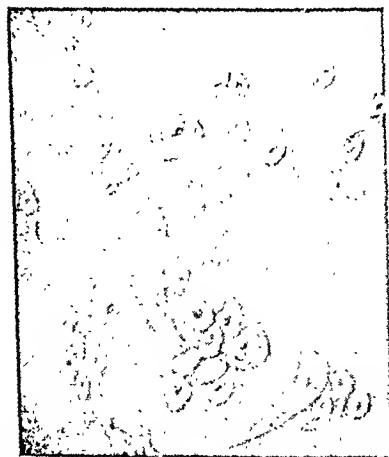
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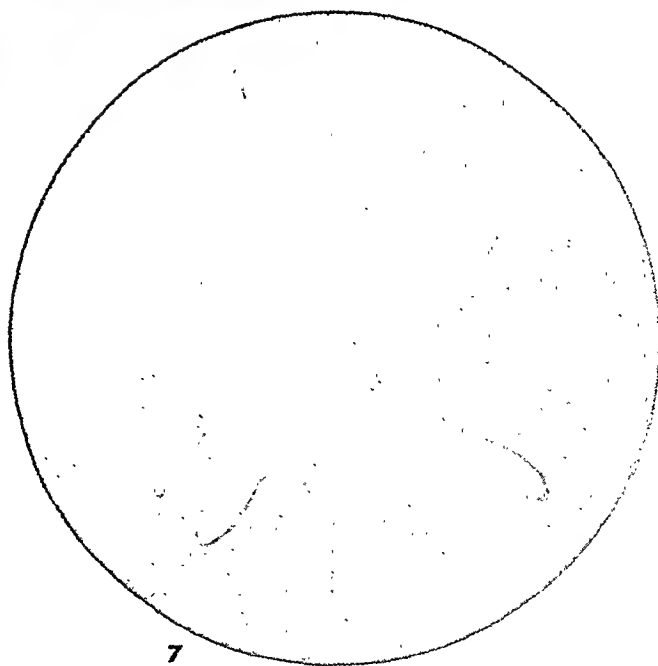
4



6



5



7

deep part of the adult nucleus there was a bright blue-green rosette composed of dense longish and round radially arranged spots which merged into one layer. The anterior Y-suture stood out densely white. In addition there was a white spotted layer of opacity in the anterior and posterior fetal nucleus. The posterior one was more prominent.

Sixth, a rosette-like branched opacity at the posterior adult nucleus with a central zone of round button-shaped opacities (nodiformis); myopia and coloboma of the macula. Chiefly on the posterior surface of the adult nucleus in the axial region there was a cloudy vacuolar opacity with whitish bands radiating to the periphery. Partly in the center, partly on the radiating strands of opacities there were about a dozen white condensations resembling buttons. Some were found on the posterior capsule. Vogt also found similar structures in spear cataract. These seem to resemble spheroliths.

Coralliform Cataract. Except for the cases reported by Riad⁵⁷⁶ and Gifford and Puntenney,⁴⁴⁸ most of the cases of this form of hereditary axial cataract in the past were described without benefit of the biomicroscope. According to the original description by Gunn⁴⁶⁵ in 1895 in a young man, 22 years of age, it was characterized by rounded or elongated opacities, the ends of which were often ampulliform. There were small crystals seen in the unaffected parts. Nettleship⁵⁵¹ described similar cases and established its hereditary nature. According to him, coralliform cataracts were made up of gray or white opacities arising from the deeper axial region and coming forward often to reach the anterior capsule. He said that each branching process expands into a trumpet-shaped opening like the "mouth of a coral" (Fig. 385). Knies (1877)⁴⁹⁸ compared the anterior projection to the vanes of a windmill. In 1906 Stephenson wrote of a case (a 30-year-old man) who had an optical iridectomy. The coralliform cataract consisted of "little pipes" in an opaque mass; some of them originated from the equator and others from the anterior surface, "the whole similar to a piece of coral." H. Fisher believed that this case represented a special form of zonular cataract.

In some instances (Riad) the opacities were elongated and resembled radiating tubes with open circular ends anteriorly. Except for some glistening crystals in the neighboring clear parts, the lens

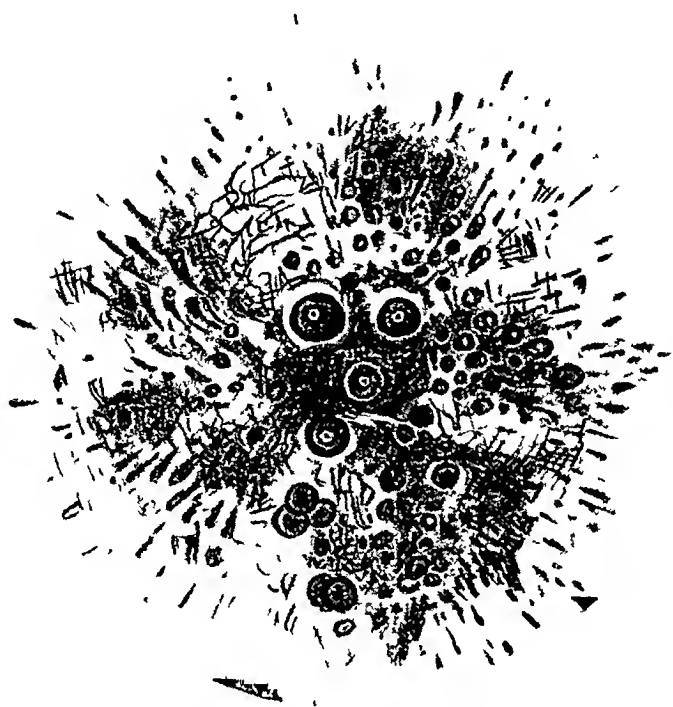


FIG. 385. Coralliform cataract.

showed no other opacities. In a case described by Gifford and Puntenney, the axial opacity was irregularly stellate or floriform and extended to the anterior capsule frontally, connected to it and facing posteriorly was a larger replica of the anterior one. In other words, in this unusual form the opacity extended through all the layers of the axial region. In these reproductions the main central lesion was surrounded by numerous white dotlike spots. These authors also reported a rare form of crystalline cataract in which the crystals were found in a compact circular mass involving successive layers of the cortex. Vogt in his discussion of spear cataract (see below) raises the point whether the so-called coralliform cataracts are related to them.

According to Gunn's picture and description this relationship is not certain. Fisher⁴²⁸ also described a coralliform cataract in which the opacity in the cortex resembled iridescent coral. An axial spindle connected the anterior and posterior cortical opacities.

Axial Fusiform Cataract. In this extremely rare anomaly the opacity extends axially through the lens thickness (Fig. 386). Hess cites two cases reported by Knies * (in brothers, in one of whom it

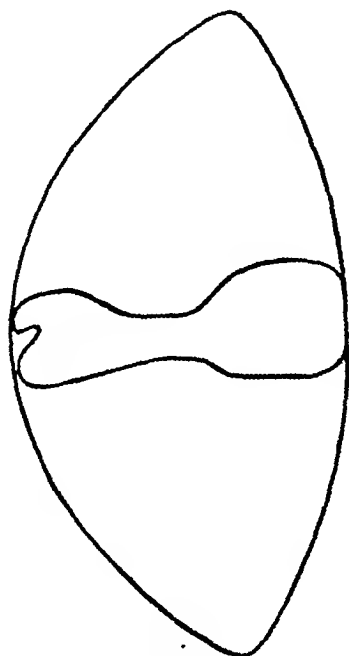


FIG. 386. Axial cataract.

was bilateral) in which apparent anterior and posterior polar cataracts were connected by a thin opacity. A possible explanation of its pathogenesis was offered by Knies and Collins (see page 1080).³⁸⁶

Spear Cataract.† This is a peculiar form of hereditary cataract, characterized by the presence of an opaque mass located in the central portions of the lens; it consists of needle-like or vermiform structures. The direction and course of these formations do not follow the normal architectural structure of the lens. The needles were frequently arranged in long bundles, a grouping often seen with tyrosine crystals. In other cases they diverged in various directions rarely crossing one another (Fig. 387). This structure, according to Vogt, differs from that of the rhomboid cholesterol crystals, which he saw in total traumatic cataract; in the latter the needles were grouped to form plates. In spear cataract it was his impression that the needles might be a product of crystallization within a shapeless mass. His first cases (1921-1922) were found in both eyes of a 9-

* I could find no other cases reported in the literature.

† Speisskatarakt (Vogt).

year-old boy whose vision was 6/24, and his mother aged 32 years. In the case of the mother there were several round button-like opacities 0.02 to 0.08 in diameter (spheroliths ?) in the central parts



FIG. 387. Spear cataract.

just in front of the posterior capsule in the right eye. The needles had spokelike projections, some of which glittered with a color display. The length of the spears varied from 0.5 to 2 mm. in length. Later examination of 42 relatives of this family revealed 10 cases of spear cataract. In some of these the opacities resembled a spinous cactus plant in that central processes had branching and crystal-like brilliant spears whose extent and direction disregarded the anatomic structure of the lens. An axial cataract was described by Gifford (1924)⁴⁴⁷ in which there were fine branching needles arranged in a manner that reminded him of two fir trees, base to base in the center with their apices approaching the anterior and posterior capsule. This could have been a form of Vogt's spear cataract.

Floriform Cataract. According to Koby, this type of rare congenital cataract, predominantly axial, seems to be an intermediate form lying between cataracta stellata and coronary cataract. Gaellemaerts, who also described a case of this kind, confirmed this conception. Other cases were reported by Veterbi and Meesmann. Koby¹⁴² stated, "the cataract is composed of a certain number of opacities, 30 to 100. Each opacity is itself formed of elements of a

round annular form. By grouping several elements, 2 to 20, there arises a series of cerulean opacities, arranged like a flower, the apparent size of which varies from 0.5 to 0.75 mm. on an average.



FIG. 388. Cataracta floriformis. (After Koby.)

The opacities are flattened occasionally like a mulberry, but sagittally have no connection among themselves. The cloudiness is more dense at the periphery than at the center and the opacities, examined by transillumination, appear diaphanous. Their color is whitish or bluish in oblique light, yellowish in reflected light. They are sometimes lustrous" (Fig. 388). These opacities are found axially in the close vicinity of the fetal Y-sutures and at times take this shape. Because of the fact that he found this cataract in one family (mother and four children, three of whom showed camptodactylia), Koby considers that the anomaly seems to behave as a dominant mendelian character. Vogt has intimated that Koby's floriform cataract resembles the ring form of cataracta pisciformis.

RARE FORMS OF CONGENITAL CATARACT AFFECTING A GREATER PORTION OF THE LENS

The cataracts referred to in this category are the diskshaped or ring form cataract and the congenital morgagnian cataract. These are rare forms of total cataract and as such have been of less interest to biomicroscopists. As a matter of fact, of the half dozen or so

reports on total cataract only in one or two recent cases have they been examined with the biomicroscope.

Disk-shaped or Ring-form Cataract. This form of total cataract is rare as a congenital manifestation. A similar or corresponding form is more frequently seen as a secondary cataract following dissection of congenital cataracts, extracapsular operations, shrunken lens and after perforating injuries (Söemmerring's ring). According to the descriptions in the literature, the congenital form appears as a thin, dense whitish opacity filling the pupil. The central part may be calcareous. In several cases it was associated with corneal opacities (leukoma adherens and staphyloma) and in one with a persistent pupillary membrane. Histologically the center of the disk is very thin and scar-like while the periphery is clubbed and composed of degenerated lens fibers, the whole formation reminding one of a umbilicated red blood cell. Ida Mann states: "The most apt embryological explanation is that the nucleus has failed to develop, has shrunk and calcified and remained adherent to the capsule so that the outer fibers could not grow around it but merely plaster themselves to the equatorial region." The reason for the failure of development of the lens nucleus is not known but it has been hazarded that the cause may lie in the failure of the epithelium lining the posterior wall of the primary vesicle to elongate to form fibers.

Congenital Morgagnian Cataract. This type of cataract is rare and is of doubtful origin. No biomicroscopic descriptions of it are available. Apparently the fault lies either in a failure of development or in a secondary degeneration of the outermost zones, since the central nucleus remains more or less intact. As a result, similar to the hypermature senile morgagnian cataract, the nucleus lies freely suspended in a fluid-like substance contained within the baglike capsule. Based on the histologic examination of such a case by Hess, Mann states: "There is not a primary failure of the outer zones but a secondary degeneration. This is shown by the fact that the lens capsule is of normal size and is filled with a milky semitransparent fluid in which the fetal nucleus floats. The outer lens fibers must have been present at some time and consequently have deliquesced."

Chapter Twenty-Five

PRESENILE AND SENILE CATARACT

PROGRESSIVE LENS OPACITIES (CATARACT) IN CHILDREN, YOUNG ADULTS, AND THE AGED

ONE should like to call some of these "adolescent cataracts," but since they appear at times in very young children when they conceivably might be congenital or later in adult life when they may be associated with frankly presenile or senile lens changes, it is difficult to name or group them according to any chronologic pattern. For example, most writers have classified them all under the heading of "presenile and senile opacities." In my opinion the inclusion of such opacities as coronary, cerulean, and dilacerated under the heading of presenile and senile, as has been done by most authors, is confusing despite the fact that some of these opacities may first appear in adult life. The use of the terms "presenile" and "senile," like that of "adult nucleus," tends to give the impression of later development than is actually the case. Starting with those preferably found in the young and eventually including all the changes which seem to be forerunners of senile cataract, and finally senile cataract itself, the following cataractous formations are listed:

1. Coronary, cerulean, and dilacerated opacities
2. Water (slits) clefts; spokes
3. Lamellary dissociation or separation
4. Peripheral cuneiform cataract
5. Various presenile and senile punctate opacities
6. Senile peripheral concentric lamellar opacities
7. Axial punctate opacities — sutures of anterior cortex
8. Immature cortical cataract (intumescent cataract)

9. Mature cataract
10. Hypermature cataract
11. Crevice formation (in mature and hypermature cataract)
12. Anterior saucer (cupuliform) cataract
13. Posterior saucer (cupuliform) cataract
14. Senile nuclear cataract
15. Nuclear cataract with double focus
16. *Cataracta brunescens et nigra*
17. Rarer forms of senile lens changes
18. Capsular opacities and folds

With age, the lens undergoes certain physical and chemical changes. It becomes relatively flatter, and the older central fibers gradually are compressed by the continual growth of the outer cortical fibers. The progressive sclerosing process within the lens (especially of the nuclei) is manifested at the end of the fifth decade by the loss of accommodation or molding effect secondary to diminution of elasticity of the capsule.* Biomicroscopically the only definite indication of these changes may be evinced as a generalized increase in relucency. The adult nuclear relief is particularly well observed in senescence. However, it is not exactly known whether this highly reflecting zone actually limits the surface of the hard nucleus, which is commonly expressed at the time of an extracapsular cataract operation. At the same time, the capsule tends to thicken and to lose elasticity with advancing years while the epithelium itself, as has been shown histologically, following the rule of senescent epithelium all over the body, involutes. It may be that some of the capsular changes (shrinkage and wrinkling processes) are demonstrated by the increasing visibility of the shagreen, already seen in young adult life, and by the rarer phenomenon, usually seen later, senile exfoliation of the zonular lamella. Duke-Elder states that "the changes in the sclerosed lens correspond to those of ageing tissues generally — a gradual dehydration with a loss of the water binding capacity, a diminished metabolism, an accumula-

* According to Vogt's idea, this would mean that the axial part of the fibers no longer are able to thicken.

tion of waste material, with the deposition of sterol and calcium deposits, a decrease in permeability, and a rearrangement of the mineral skeleton of the tissues." These are the physicochemical manifestations of lens senility, inherently and genetically determined, as are all the changes of senescence governed by species, individual, and even organ determinants.

The complicated question arises, if, when, and how much are the changes of senescence influenced by the so-called "exogenous" influences, such as toxins arising from fatigue; radiant energy; endocrine dyscrasias; nutritional deficiencies; and infections (see discussion on diabetic cataract, p. 1177). Also, how much of a role do these play, for example, in the presenile lens changes? Are they the result of an inherent abiotrophy (premature senility), some of which so closely resemble cataractous changes seen in the frank endocrine cataracts? These questions are as yet unanswerable but at the same time certain definite morphologic patterns do occur in presenile and senile cataract (as well as in congenital cataract) in regard to types and specific localization of the changes. Vogt with considerable evidence has made a strong plea for heredity, suggesting that not only the lens as a whole, but even groups or sections of fibers (e.g., cortex or nucleus) have individual specific genetic determinants which when faulty could lead to dissolution or weakness. In this way also, outside influences (environmental) could secondarily precipitate cataractous changes easily. Vogt has shown, especially in his studies of identical twins, that the appearance of such changes are chronologically as well as morphologically similar in spite of difference of environment. The finality of such a conception, although true within limits, if carried to the extreme, could easily result in a state of defeatism — a condition not unlike that existing in the eras of antiquity and the middle ages when investigators were satisfied with authoritarian systems of identification and classification only. As it now stands, we must await the physicochemical advances of the future because it may be shown that what some investigators now consider inexorable laws of nature may be in the end controllable.

It will be seen that Vogt does not clearly separate presenile lens opacities from those that are definitely known to form senile cataracts. For example, in cases in which presenile opacities (coronary)

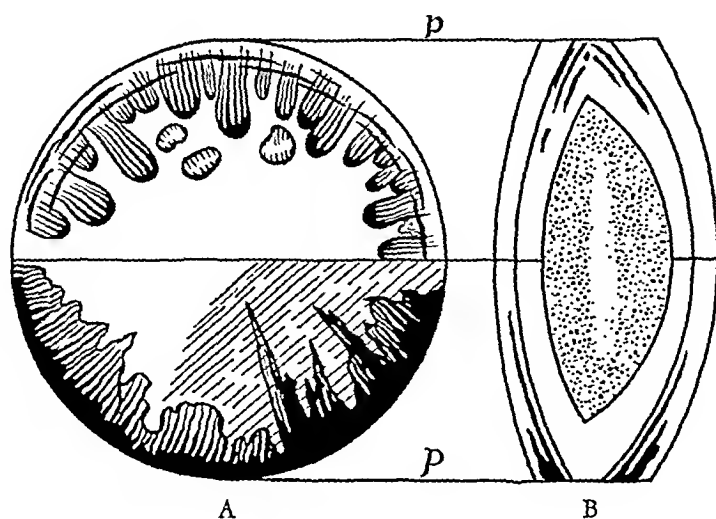


FIG. 389. Diagram showing the principal types of opacification of the lens. A. Frontal view. B. Optic section; *p* marks the border of the dilated pupil. Above, opacities, chiefly club-shaped, of a coronary cataract; below, senile cuneiform opacities. In A, to the right, lamellar separation. In B, in the middle, nuclear cataract involving only the fetal nucleus and leaving free a little central interval.

are found in the second and third decade, water slits, lamellary separation, and spokes frequently will form so that after a time an eventual cortical cataract develops. This association or gradual transition of these forms is not uncommon. However, it should be pointed out that certain of these so-called "presenile" opacities are observed in the young and, as such, may be confused with congenital cataracts. Most of them are found outside of the region of the inner fetal nucleus and their Y-sutures or in between the central coffee beanlike nuclei and the stripe of the adult nucleus and in the cortex. The deeper part of the so-called "adult" nucleus is already present at birth and separates the capsule from the inner fetal nuclei. In addition, except for cataracta dilacerata and some isolated small flat opacities, the presenile opacities, particularly those found in the young, are chiefly peripheral (e.g., coronary and cerulean cataracts). It is interesting to note that nuclear cataract begins in the fetal parts of the lens (Fig. 389).

CORONARY CATARACT (CATARACTA CORONARIA; CERULEAN
CATARACT; CATARACTA CERULLA [BLUE]
ET VIRIDIS [GREEN])

The biomicroscope demonstrates easily that this is one of the most common forms of presenile cataract but because of its peripheral location behind the iris it may be overlooked unless the pupil is dilated widely. According to Vogt (confirmed by Gjessing⁴⁵¹) and others * cataracts coronaria et cerulea are found in at least 25 per cent of all persons after puberty and are hereditary. Usually bilateral, they may appear to be stationary but over long periods of time, most of them progress slightly to a varying degree. This is seen in cases observed over a long period of time. In the early stages the opacities are isolated or few in number but later they tend to develop into a continuous wreath. As a rule, by themselves they do not disturb central vision unless associated with other types of lens change, especially senile cataractous alterations. Coronary cataract begins in the periphery, occupying that portion of the lens corresponding to the border between the middle and outer third of the lens radius, appearing very much like a crown or wreath situated on the equator of the adult nuclear zone. Frequently the opacification extends within the adult nuclear stripe or external to it within the deeper cortex. The surfaces of the opacities form a thin layer concentric to that of the adult nucleus and according to Vogt extend onion-like. This corresponds to the direction of the concentric lamellae (page 1010) seen so distinctly in macerated preparations and which, according to him, are not related to the radial lamellae of Rabl. The shape of the opacities vary (clubs, disks, lines and rings) but the most prominent are club-shaped. The thickened sharply outlined, rounded parts of the clubs extend in an axial direction reminiscent of dentate stalactites. Peripherally the tapered processes of the clubs may extend posteriorly around the equator of the adult nucleus or may terminate at the equator in a diffuse manner (Plate LXVI). Frontally finer radiating linear or spiral opacities

¹ Weissenbach, 1917;⁶⁷⁴ Krenger, 1918;⁵¹⁴ Horlacher, 1918;⁴⁸⁰ Kirby, 1936.⁴⁹⁴

PLATE LXVI

FIG. 1. Coronary cataract. Diffuse illumination.

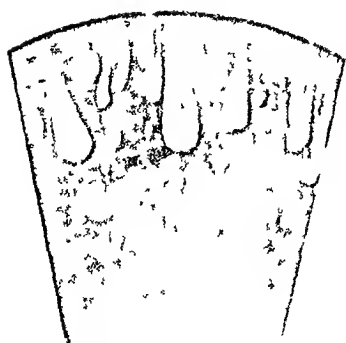
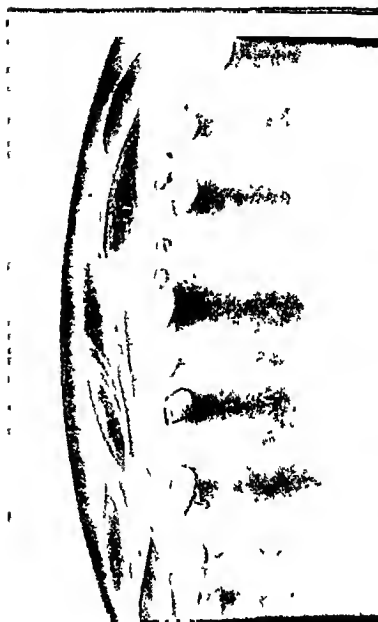
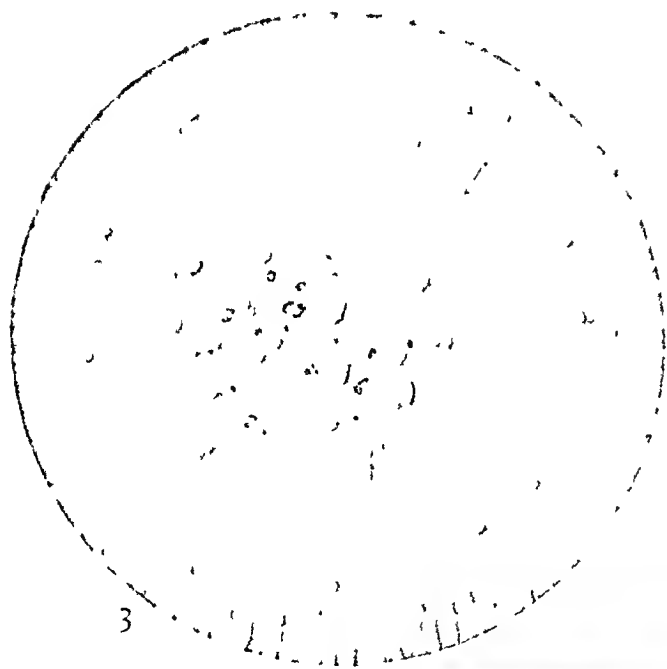
FIG. 2. Same case as shown in Figure 1. Direct focal illumination.

FIG. 3. Coronary and cerulean cataract. The central opacities exhibited a vivid color display. Note signs of cortical changes. (Water clefts and lamellary separation). Diffuse illumination.

FIG. 4. Same as case in Figure 3. Direct focal illumination.

FIG. 5. Coronary and cerulean opacities showing peripheral clubs and smaller rounded forms axially. Diffuse illumination.

FIG. 6. Same as case shown in Figure 5. Direct focal illumination. High power.



are frequently seen. The linear appearance springs from the fact that observation is directed at the edges of the opacity tangentially as they extend around the equator. In some cases the clubbed opacities may be so close to one another as to seemingly fuse. The color of these opacities varies according to their density. Thin forms may appear grayish with a faint bluish tint while the well-developed ones are whitish. No exact explanation for the formation of these clubs has yet been given, but Vogt has suggested that perhaps it is related to the expansion of fluid between the concentric lamellae of the lens. In a case of traumatic cataract, he has seen that vacuoles (shaped like clubs) can lead to opacification. If the vacuole is located peripherally, it becomes a club-shaped opacity, and if it is more axial, it assumes a discoid or ring shape.

In addition to the characteristic clublike formations, more axially located discoid or ring forms may be found. They begin as very thin grayish or bluish nebulous opacities in the deeper cortex, and are only visible by direct focal light. Later when these roundish opacities tend to become more opaque and white they may be observed by retro-illumination.* They may extend centrally but because of their transparency do not usually affect vision. However, exceptions occur in which, owing to increasing density, these opacities, when axially located, may interfere with vision. Frequently irregular groups of dotlike opacities may be seen between the disks and rings. Another typical form of opacity, usually associated with the clubs are the linear concentric opacities. They appear as broken

* The color variations seen in these opacities probably depend on their screening action of incident light. As in blue irides the coloration manifested by the semitransparent whitish iris tissue when viewed against the dark background of the posterior surface is a consequence of the diffusion of incident light so the cerulean opacity gains its color as it is viewed against the dark pupillary background. Depending upon the degree of light diffusion within the opacity (nitra lamp) a greenish hue may be imparted (cataracta viridis). Frequently retro-illuminated light from the deeper portions of the lens (mirror reflex) may give anteriorly located opacities a brownish hue. The thicker opacities appear white in direct focal illumination. However, the fact that coronary and cerulean disk- and ring-shaped opacities may show various spectral colorations (from bright red to violet) has caused many to question the origin of this phenomenon. As an analogy Vogt cites the work of Ehrenhaft (1914) and of Gerde Laski (1917) which indicates that the color of small colloidal molecules depends upon their size. In this way the smallest appears violet while the largest appears red. Intermediary ones would be green or blue. This hypothesis of color dispersion was also referred to earlier by Hess (1911).⁴⁷² According to Bellows, Hess' explanation is based on the observation of Lord Rayleigh who found that in an opalescent medium containing innumerable particles of a different refractive index, the dispersion of light is inversely proportional to the fourth power of its wave length.

or dashlike concentric lines running at right angles to the direction of the clubs and are situated in the extreme periphery. To see them it is often necessary to employ a wide angle of observation and illumination, comparable to that used when looking at the periphery of the fundus with the ophthalmoscope. The direction of these short concentric lines is not unlike the longer and perhaps slightly more axially located stripes in the deep cortex in cuneiform cataract. As a matter of fact, coronary opacities (especially in adult life) may be associated with the latter as well as many other types of presenile opacities (e.g., dilacerated forms). The formation of water slits leading to spokes (which appear thin in sagittal section)* in cases of coronary cataract is a concomitant senescent finding. Vogt stated that this combination is hereditary and is one of the most frequent of all presenile lens changes, and that after one learns to recognize these characteristic thin spokes, which often extend into the axial region, the presence of a coronary cataract may be predicted, even before dilating the pupil.

Cerulean cataract is often found together with coronary opacities. The former, however, is distinguished by the presence of discoid, ringlike or irregularly shaped opacities, colored blue or green and limited to the adult nuclear region. This type of cataract, which undoubtedly is a subform of coronary cataract, is found in younger individuals (Plate LXVI, fig. 5). Consequently in some instances it may represent a congenital form. In other cases the opacities may be situated on the surface of the adult nucleus, so it is difficult to make any definite statement regarding their true time of origin. Cerulean opacities have even been identified in hypermature cataracts (Vogt). The fact that the blue color is retained in spite of the white background is an exception to the previous explanation of the color changes. However, in these instances, the presence of a layer of less reluctant fluid may contribute to color formation.

This type of colored opacity may be seen rarely in the inner fetal nucleus, so that at times a peripheral coronary cataract may be associated with central opacities of the type seen in cerulean cataract.

* These spokelike or coronary opacities may be very thin and frequently show coloration

Coronary and cerulean opacities may also occur concomitantly with many of the developmental cataracts referred to before, e.g., stellate, anterior, axial, embryonal cataract, etc. The direction and location of the smaller bluish dotlike, or at times irregular or spiral threadlike opacities vary from case to case. In the periphery they may form concentric layers, while those situated more centrally may be arranged radially. Despite the circumscribed opacities found within the nucleus it is usually less relucant than the cortex, giving the impression of a dark area surrounded by a hazy cortex. The short spiral radial lines, which were mentioned before as located at the equator between the clubs, can also be found more axially. Vogt gives their average size as from 0.12 to 0.2 mm. long and 20 μ thick. Their direction and shape seem to correspond to that of the wavy sclerosed nuclear fibers. They may appear brownish or grayish in color.

Although coronary cataracts are generally considered to form at the time of puberty, the fact that analogous forms of thin color-displaying spots may be found in the deeper nuclei, and that they are bilateral and familial in occurrence, leads to the conclusion that these slightly progressive forms are all related and are genetically determined.

DILACERATED CATARACT (VOGT) (CATARACTA DILACERATA —
MOSS-LIKE CATARACT)

This type of opacity, first described by Vogt in 1921 is found frequently in association with coronary, cerulean, and centrally located congenital cataracts. Morphologically it resembles a small flat piece of teased-out moss or sponge with frayed edges (Fig. 390). Similar to the linear coronary opacity of the periphery, the opacities tend to extend along a lamella in a concentric direction (Fig. 375 B, C). The fact that opacities do extend in this way seems to substantiate the idea that the lens is composed of concentric lamellae as well as radially directed fibers. The color of dilacerated opacities is usually gray. However, occasional whiter-looking strands may be

seen. Also like coronary and cerulean opacities a dilacerated opacity may reveal a yellow, blue or green coloration.

In an individual lens these opacities may appear singly or multiple.



FIG. 390. Dilacerated cataract.

Dilacerated cataracts may be within the adult nucleus but not necessarily in the axial regions. In one case, in a man who had bilateral coronary cataracts I saw four of these thin grayish opacities, in one eye only, arranged circularly just below the surface of the adult nucleus. They were located in an area corresponding to the middle third of the lens radius. One of them had a definite leaflike ribbed internal structure. The main part of the opacities was composed of fine irregular threads and dots. The immediate area surrounding it was hazy, gradually fading into the background. Vogt said that he was unable to determine at what age dilacerated cataracts developed, but that they were probably congenital or acquired early in life. It is not known whether these opacities are stationary or progressive.

SENILE CATARACT

Under this heading we shall first consider the progressive types of presenile and senile change that morphologically seem to be the underlying basis of or at least an integral part of the lesion known clinically as "senile cataract." Because these changes (e.g., punctate dots, water slits, spokes, lamellary separation, cuneiform opacities, and nuclear opacification) may appear early in adult life and may

span decades before they seriously interfere with vision, they have been classified as presenile. But rapid extension of some of these processes at any time may result in the appearance of a mature senile



FIG. 391. Mature senile cataract. Frontal view showing large radial water slits.

cataract within a very short period. Before the days of biomicroscopy such changes, when sufficiently advanced to produce opacities visible with the loupe or ophthalmoscope, were classified according to their location as cortical, nuclear, and posterior lamellar cataracts; and according to their development (and incidentally their effect on vision) as early or incipient, intumescent, mature, shrinking or hypermature, and morgagnian or liquefied cortical cataracts. Nuclear cataracts were classified as early or well-developed with subdivisions such as central hyperrefringens (for lens with double focus [Koby]) and cataracta brunescens and nigra when accompanied by color changes. However, even though biomicroscopy has advanced our knowledge concerning the morphologic development of these opacities, at the present time it has warranted no change in the older clinical classifications or terminology.

It is a common daily experience to find cases in which all types of changes (cortical, nuclear, and posterior saucer) are simulta-

neously present. With the narrow beam, even in these cases the separate lesions can be distinguished except, of course, when the cortex is coagulated and opaque. As already discussed, the cortex is composed of younger fibers and is located just below the capsule; it reacts morphologically in a different way than the nucleus, the fibers of which are older and more compressed. These differences are apparently due to physiologic sclerosis of the nucleus.* This process is protective since it retards rapid or intense opacification thereby conserving vision for a longer time. The water binding power of the lens varies with age so that with increasing "sclerosis" the water content increases. We are unaware of this process biomicroscopically until proteolytic enzymes causes disintegration and consequent opacification. In discussing the chronologic † progression of the presenile and senile changes leading eventually to the clinical forms of senile cataract, Vogt tends to disregard the local metabolic and physicochemical influence preferring the genetic theory. He considered these alterations so far as rate of development and time of inception is concerned as being comparable to those seen in other tissues, e.g., senile atrophy of the pupillary margin, arcus senilis, graying of the hair, etc. Vogt stated that he could prove that more than 90 per cent of persons over 60 years of age have senile opacities of the lens varying individually in the same way as senile changes in other organs. Moreover, if the question is asked why all lenses do not become completely opaque with age, then the same question must be asked concerning time of appearance of other senile changes.

From this standpoint the difference in behavior of the cortex and the nucleus is manifested by the greater ease with which the sutures and fibers of the cortex become separated by fluid, eventually resulting in an intense opacification (precipitation of lens protein?). The extreme is seen in morgagnian cataract in which complete dissolution and liquefaction of the cortex occurs. The nucleus seems to react less violently than the cortex. It simply becomes more or less uniformly cloudy.

* An excellent summary of present day knowledge of the composition and metabolism of the lens is given by Bellows.³⁶⁴

† Since in some cases these changes develop rapidly within a few months and in others are stationary for years.

CORTICAL CATARACT

In the cortex especially, the biomicroscope permits visualization from the very beginning of gross changes that apparently are associated with the imbibition of water, and the degeneration of the lens fibers. The earliest manifestations of these cortical alterations are the formation of water clefts (slits), lamellary separation, and groups of punctate opacities, probably similar or related to punctate opacities of the central cortical sutures. The opacification attending these processes go to make up the so-called incipient cataractous alterations of the cortex. Clouding of the fibers of the walls and contents of water slits results in "spoke" formation. Separation of the concentric lamellae leads to peripheral cuneiform (pyramidal) opacities. Further extension of all these processes results not only in progressive opacification but also in swelling or intumescence.* Maturity is marked by almost complete opacification of the cortex. From this point on, owing to dialysis of water and disintegrated lens substance, regressive changes (hypermaturity) follow, characterized by shrinking. The rate of progress of these changes varies greatly, sometimes extending over years, with alternating periods of arrest and progression; in others cessation of development may occur at an early stage, resulting in an arrested form of incipient cataract. Loss of vision in these cases, particularly in the aged, may rather result from nuclear clouding than from cortical changes. On the other hand the progression to maturity of cortical changes may proceed with astonishing rapidity.

WATER CLEFTS (SLITS) AND SPOKES

Water clefts, a manifestation of the absorption of water by the cortex, are characterized by the appearance of dark spaces (of lesser optical density) in the form of (1) radially directed sagittal slits,

* Bellows, citing Krause's (1934)⁵¹³ hypothesis on the fluid "traffic" in cataract, states: "Following injury to the lens, acidosis results which stimulates certain proteolytic enzymes to act upon the lens substance. The fragmentation of the lens protein yields many smaller particles, increasing appreciably the osmotic pressure within the capsule. Water is accordingly attracted into the lens and the lens swells. In a rapidly forming cataract the end products of these hydrolytic processes accumulate, and marked swelling results. As the action of the enzymes continues, the protein residues are broken down to crystalloid size and diffuse out of the lens."

PLATE LXVII

FIG. 1. Water clefts in the anterior cortex. Direct focal illumination.

FIG. 2. Water clefts in the posterior adult nucleus (girl, 16 yrs.). Direct focal illumination.

FIG. 3. Spoke formation as seen by retro-illumination.

FIG. 4. Deposits of myelin droplets within a spoke, as seen by retro-illumination.

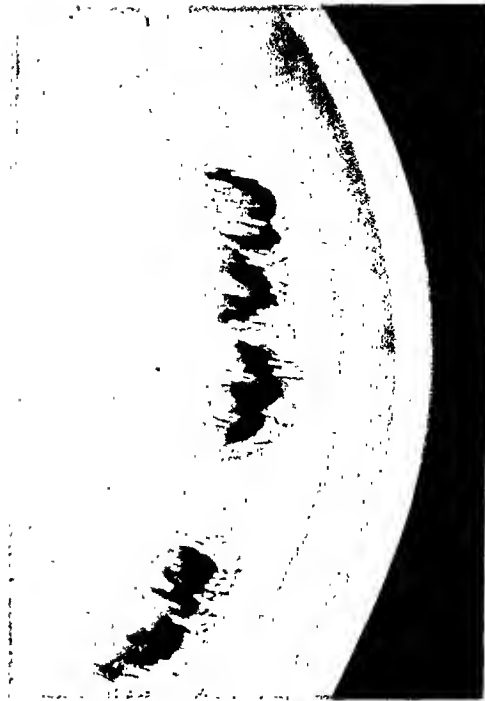
FIG. 5. Droplets seen in light reflected from the fundus.

FIG. 6. Lamellary separation associated with pyramidal or cuneiform opacities.

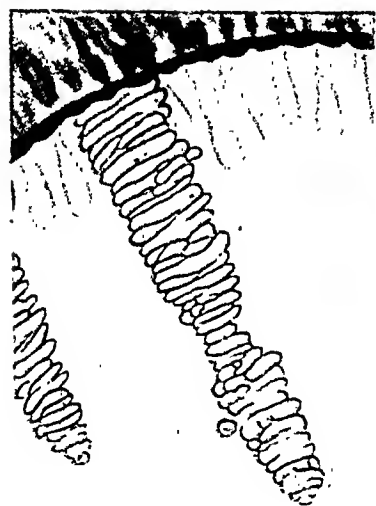
FIG. 7. High-power view. Direct focal illumination. Optic section showing details of a water cleft and laminary separation.



1



2



3



5



4



6



7

corresponding to the sutures or (2) in larger lamella-like separations (corresponding to the concentric lamella) which extend sheetlike frontally. Water slits may form also as a result of separation of fiber bundles. This type has also been noted in traumatic cataract and in massage cataracts. In the latter cases, particularly in the aged, massage of the anterior lens capsule without rupturing the capsule, can cause a localized disruption of lens fibers. Spokes, so commonly seen in the periphery of the cortex, are water slits with partial or total opaque contents.

Earlier authors (prebiomicroscopic) described radial slits and spokes and surmised that they were fluid spaces but it was Vogt (1919-31) who showed their connection to the sutures and who also demonstrated the larger fluid spaces caused by separation of the concentric lamellae. According to him, the latter confirms the idea that the lens is made up of layers of fibers which are concentric as well as radial.

Radial water slits may be seen in a more or less hazy way, frontally (especially in intumescent cataracts) by oblique or diffuse illumination and the peripheral ones, i.e., those associated with spoke formation, can be seen ophthalmoscopically as clear spaces between spokes provided the pupil is dilated. Owing to retroreflected light from the deeper portions of the lens, water slits may appear blurred especially when employing the wide biomicroscopic beam. In order to see water slits clearly, especially in cross-section, the narrow beam is essential (Plate LXVII, figs. 1, 2, 7). This is particularly true for water slits of the posterior cortical layers, where owing to diffuse reflection from the anterior portions of the lens, they can only be seen with the optic section (Fig. 392). With this method the appearance of the radial slits is striking. Dark, irregular, but sharply defined areas are seen outlined within the gray lens substance. Slight lateral movements of the beam will give "serial" sections through the slit and usually disclose its irregular shape and extent. The distinctness of water slits and of lamellar separations increases as the zones of specular reflection are approached. Sudden narrowings of the lumen of the slits are common as are sharp changes in direction. This at

times tends to give the water slit a jagged appearance (Plate LXVII, fig. 1). The irregularity of the walls causes them to have sharp splinter-like processes which protrude into the lumen of the slit.



FIG. 392. Water slit seen by optic section. Subcapsular vacuoles. Vacuoles on the surface of the adult nucleus. Peripheral spoke in cortex below.

This jagged appearance may be caused by the separation and breaking up of fiber bundles. In other words, instead of flat radial spaces separating the sutures (as seen frontally by diffuse illumination) the optic section will disclose that the water slits have a definite sagittal thickness which may extend irregularly through the width of the cortex from the surface of the adult nucleus to the line of disjunction. In only very rare instances do they project through the line of disjunction to reach the capsule.* Their common location is in the middle and deeper parts of the cortex. Likewise it is extremely rare to find a water slit within the adult nucleus, although I did see several in a single case of traumatic intumescent cataract in a 16-year-old girl in which the slits seemed to extend into the adult nucleus. In the frontal view as well as in optic section it will often be noticed that slits result from dilations or separations of portions of the sutures

and, corresponding to their direction, radiate from the axis. The ends of the slit are usually pointed toward the intact part of the suture and often empty into it. At times one slit runs into another at the place of branching of the suture or a clear slit may empty peripherally into an opaque slit (spoke). In addition delicate white fibrils, concentrically directed, may frequently bridge across a dark tunnelled-out water slit (Plate LXVII, fig. 7). These are more commonly seen than radially directed bridges. Vogt believes that this proves a stronger continuity in radial fibers than in those concentrically directed. Macerated preparations show that the bind-

* According to Vogt, directly subcapsular slits are very rare and similar to other subcapsular opacities (e. g., saucer cataract) indicate a poor prognosis; since in such cases opacification progresses quickly. Degeneration just beneath the capsule shows that the youngest lens fibers are partaking in the process.

ing together of lens fibers in the direction of Rabl's lamellae is more solid than those of the concentric zones. By diffuse illumination it will be generally noticed that the areas (walls) contiguous to the direction of the slit are more relucant in certain places. This opacification has been explained as being brought about by the imbibition of fluid by the fibers nearest to the water slit. As a result the borders of the slit may appear partially as a narrow whitish line or even as radiating broader bands. In some cases this increase of reflection may be seen in optic section as outlining the jagged irregular edges of the slit. Since such opacification is seen commonly in intumescent cataract (water slits are an underlying process of intumescent cataract, whether senile, toxic, or traumatic), it may be an indication of rapid progression.

Becker³⁵⁹ noted that dark water slits may appear opaque on change in direction of the incident light. This appearance is probably caused by regular (specular) reflection, not unlike the whitish color of the shagreen of the capsule seen in normal lenses. The shagreen itself when viewed over a water slit appears darker than otherwise even when the cleft is located deep in the cortex. This unexpected phenomenon may be caused, as suggested by Vogt, from lack of reflection of the subcapsular concentric layers unless they are interrupted by the formation of a water slit. Ordinarily brightness of the shagreen is augmented by diffuse and regular reflections from these layers. Butler, basing his contention on the point that he could see a capsule shagreen in morgagnian cataract (when the cortical fibers beneath the capsule were liquefied) believed that the shagreen results from specular reflection of the hyaline capsule alone. Von Hess concluded that the darkening of the shagreen in front of the clear water slits proves that the location of the latter is just below the capsule.

Using superimposed glass slides Vogt demonstrated experimentally the reason for the darkening of the shagreen over a water slit; he also demonstrated the optical illusion whereby in diffuse illumination they appear to be more superficial than is actually the case and the reason for apparent opacification of clear slits seen on alternating

the direction of illumination and observation.* The fact that water slits are difficult to see at times and readjustment of the angle between observation and illumination is required, demonstrates, according to

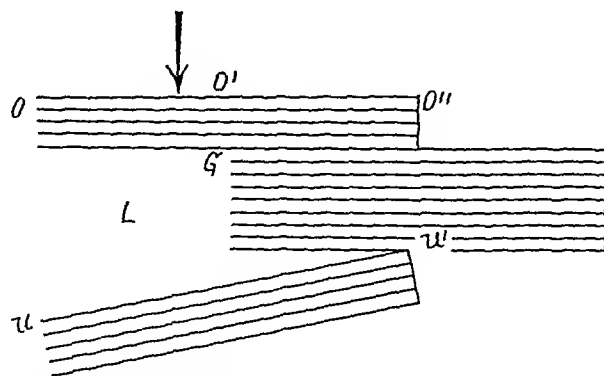


FIG. 393. Vogt's arrangement of glass slides to illustrate optical phenomena of layers of cortex.

Vogt, that their refractive index differs little from that of the cortical substance. This is also the reason for the weak reflection of their walls. Their dull asbestos-like relucency becomes visible only by a certain angulation of the beam's direction. The index of refraction of the dark water slits is comparable to that of the corneal epithelium, the aqueous, or the fluid parts of the vitreous. Consequently Vogt reasoned that the fluid within the slits is derived from the aqueous and that this fluid differs from that of vacuoles. The latter have a different index of refraction, and hence are more easily seen because of their vivid glitter; in addition they are more readily visible in retro-illumination than are clear water slits. According to recent investigations, vacuoles consist of "myelin" droplets, a degenerative product derived from the substance of the lens fibers;

* Vogt arranged a series of glass slides (Fig. 393), one over the other to represent the layers of the cortex. The middle ones were pushed out a little so that a gap formed, *L*, which might be compared to a water slit. The deepest layer was placed obliquely, *U.U'*, in order to imitate the irregular direction of the walls of a water slit. The whole arrangement was then placed on a dark background so that *L* appears as a dark gap. A film of vaseline was applied to the upper surface *O—O''* to make it uneven and to simulate a shagreen field. If light of low intensity is permitted to reflect from this upper surface, the dark gap *L* is still seen in spite of it. The reflection from *OO'* is less intense than from *O'O''*. Also as a consequence of refraction the location of the gap *L* seems to just be under the surface *OO'*. In other words as a result of refraction the gap suffers an apparent dislocation toward the surface. With a wider angle of observation it appears as if *L* shifts toward *OO'*. Such a shifting *in vivo* could easily result in an incorrect localization so that a deep cortical water slit might be thought to lie subcapsularly. If the light is only allowed to be reflected from *U.U'*, then *OO'* appears brighter (relucient) than *O'O''*. This could explain why clear water slits appear "opaque" in certain directions of illumination and observation.

biochemically myelin (a bi-refrangent substance related to cholesterol) is insoluble in water or alcohol.

Investigating the frequency of water slits, Pfeiffer⁵⁶² in Vogt's clinic found that in 84 persons over 50 years of age their incidence was about 36 per cent and that they were twice as frequent in the anterior cortex as in the posterior. Koby states that "clear clefts appear after 50 years of age and it seems a little earlier in women than in men." Kirby⁴⁹⁷ reported the presence of water slits and lamellary separation, alone or together, in about 12 per cent of 945 cataracts. Vogt has stressed the frequency of water slits in association with progressive coronary opacities in individuals between 30 and 60 years of age. In many of these cases the progress was extremely slow extending over decades and in spite of spoke formation vision may be only slightly affected if at all. In addition in some of these cases he was able to observe the hereditary nature of these coronary spokelike cataracts. According to Duverger and Veeth in 10 per cent of all cases seen, water clefts were observed in the absence of cataract or without cataract developing later.

Spokes. The opacification of radial water slits results in the formation of spokes which probably are one of the most common forms of lens opacities (Fig. 391). This is a further extension of the process which in some cases proceeds slowly over decades and in others (when associated with extensive lamellary degeneration, fluid imbibition, and eventually individual fiber decomposition) leads to intumescent cataract. It is not unusual to find cases in which a few peripheral spokes develop and upon the formation of a nuclear cataract, the degeneration in the cortex is arrested, so that ultimately the loss of vision is due more to changes in the nucleus than to those in the cortex. Nuclear cataract in these cases would almost appear to inhibit the advance of cortical changes. Although we would like to think that only the central parts of the lens (adult and fetal nucleus) are subjected to sclerosis with age, evidently to a lesser degree this hardening can occur in the older parts of the cortex. When this happens it could easily inhibit the progression of spokes.

The opacification of the radial water slits with resultant spoke

formation is a consequence of the deposition of myelin. This substance apparently is not soluble but rather forms a cloudy emulsion within the slit. This process is also seen to occur more rapidly in

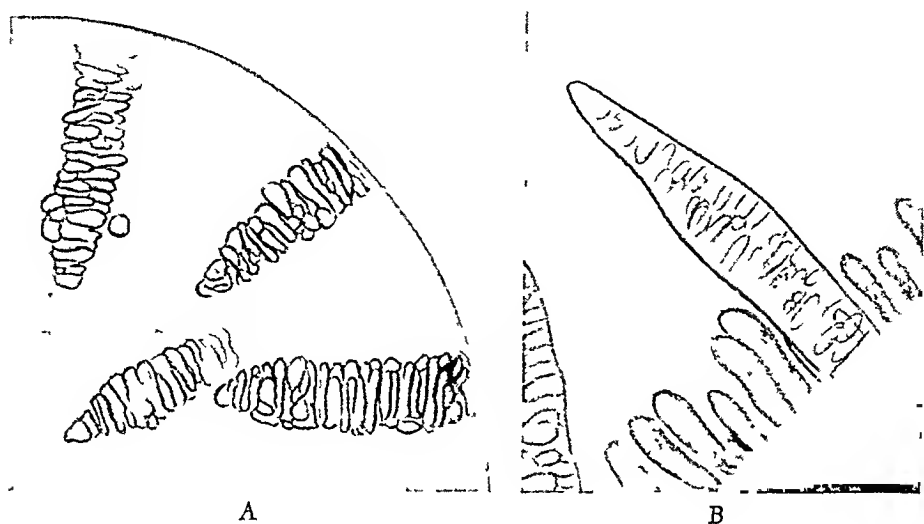


FIG. 394. A. Spoke by retro-illumination. B. Spoke with myelin droplets viewed by light reflected from the fundus. High power.

intumescent cataracts where not only the sutures and lamellae are separated by fluid and myelin but finally also the individual lens fibers themselves. Such a reaction can be reproduced within a day or so in macerated preparations.* Ordinarily the conversion of a clear water slit into an opaque spoke may take several years. With higher powers, myelin droplets appear as whitish opaque bodies but by retro-illumination their vacuolar nature becomes apparent (Fig. 394 A, B). Clear water slits cannot be seen, but vacuoles (sub-capsular or within water slits) have a yellowish tinge, especially if viewed in the light reflected from the posterior mirror reflex (Plate LXVII, figs. 3, 4). Depending on the direction of the light the edges nearest the light will glow (unreversed illumination). (See Vol. I, page 90.) In addition, lens vacuoles, similar to those in the cornea, will project shadows deeper when viewed by direct focal light. Depending on the refractive index of the vacuole, as compared to

* In these preparations, a freshly removed lens is subjected to the maceration effects of a watery solution. Upon removal of the capsule sectors of lens can be easily separated by splitting of the sutures. In this way not only can the hydration changes comparable to senile changes be observed in the lens but also the concentric laminations of the sectors.

that of its surroundings, three refractile phenomena may occur (Koby): (1) If the refractive index of the contents is greater than that of the surrounding media, it acts like a convergent lens, producing a luminous cone bordered by two dark shadowy bands. (2) If the refractive index of the contents is less than that of the media, a divergent lens action results. This leads to a projected shadow of the vacuole encircled by a more or less divergent luminous beam. (3) When the refractive index of the body is the same as that of the media, a direct shadow formation occurs. (See Vol. I, Fig. 105.)

In light reflected from the fundus the myelin vacuoles within the spoke appear red (page 981). (See also, Fig. 394 B; Plate LXVI, fig. 5.) As opacification of the contents of the water slits progresses the walls become increasingly opaque, evidently due to deposits of myelin in or between the adjoining fibers, possibly combined with pressure effects. The opacification of the walls in a radial direction causes the appearance of spokes or riders so commonly seen in the peripheral parts of the cortex ophthalmoscopically. However, some of them may result from cuneiform opacities (see below). In direct focal light the age of a spoke is determined by its color. Old spokes are distinguished by their vivid opaque whiteness.

Several authors, Stanka,⁶¹⁶ Galla, Klainguti⁴⁹⁵ and Vogt,⁶⁵⁵ have demonstrated after iridectomy that movement of vacuoles and other cortical opacities occurs with accommodative effort. Opacities move axialward during accommodation and then return to their original peripheral location during rest. Vogt believes that this represents proof of the intracapsular mechanism of accommodation (page 1016). He showed this in a case of hypermature cataract in which radially directed opaque fibers bent during accommodation and resumed their original shape on rest.

Lamellary Separation (dissociatio lamellosa) is characterized by the presence of more or less parallel lines in the visible and deep cortex. They are generally found in connection with a sagittal separation (evidenced by dark spaces) of the so-called cortical concentric layers (Plate LXVII, fig. 7). This is a second type of water slit. Like the radial water slit, it is intimately associated with cata-

ractous change of the cortex; it is also seen in macerated preparations. Time of appearance and rate of progression varies greatly from case to case. Whereas the formation of radial water slits repre-

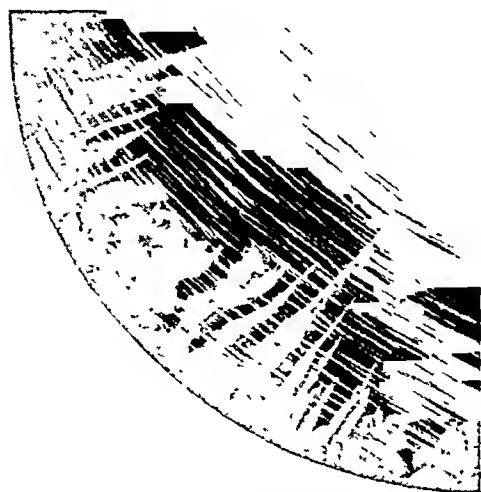


FIG. 395. Lamellary separation, frontal view. Nasally, the separated fibers are seen passing over a cuneiform opacity.

sents the changes governed morphologically by the radial direction of the lens structures, lamellary separation is the expression of the same changes of the so-called concentrically directed layers (Vogt). Just as spokes develop from the opacification of the radial water slits, so do the peripheral cuneiform opacities arise from opacifications of the concentrically separated lamellae. (Fig. 395). There are still a great many unsolved problems concerning the nature of lamellary separation.

Owing to hydration, great pressure strains are created within the lens. This serves to separate along the lines of least resistance and according to their direction the various components of the lens structure. Evidently the sutures (radial) and the so-called concentric lamellae first show the effect of this stress, not unlike the opening of the seams in a leaky boat. Despite the fact that the lens fibers (Rabl) in equatorial section form radially directed lamellae, macerated preparations show a tendency toward concentric separation (see Fig. 367, p. 1011). The latter, although not actually representing the true morphologic direction of the lens structure, evidently indicates that strains can cause separations in this direction. It is

difficult to explain this phenomenon except by the theory that fibers of the same age, being chemically similar and under strain, can separate sagittally and in a frontal way and as a result give the im-

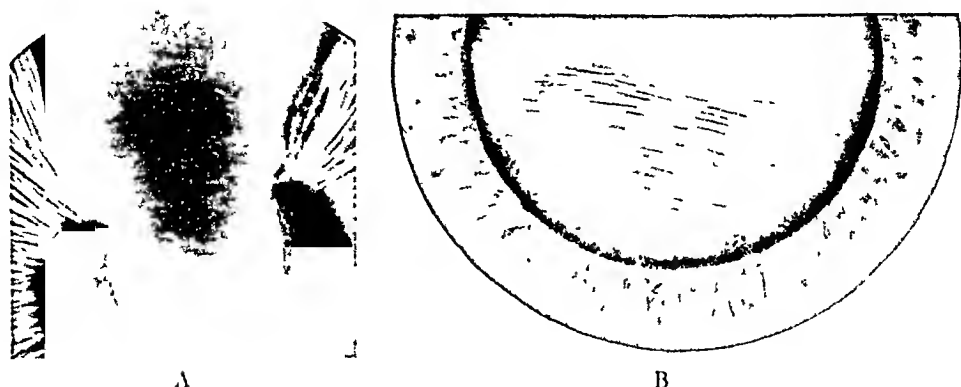


FIG. 396 A. Form of lamellary separation. Parallel lines of separation extending across the lens without regard to suture system. B. Parallel lines of lamellary separation extending across the lens in front of cuneiform opacities.

pression that the lens is made up of concentric layers. In other words, here we are confronted by the fact that separations can occur ("across the grain") which do not apparently correspond to the normal directions of the radially directed layers. In 1912 Vogt saw these lines in the deep cortex and at first thought that they represented fine folds (Fig. 396). These parallel lines can be seen in their full extent at the level of the nuclear relief by somewhat diffuse illumination when the pupil is dilated. It was found that they are more commonly located in the lower nasal sector. Predominantly, their line of direction is steep, starting from below temporally and running upward nasally. Simultaneously when this direction of lines occurs, cuneiform opacities will frequently be found nasally and below (Fig. 395). Since these lines and intervening gaps may extend in other directions, Vogt pointed out that it is more proper, strictly speaking, to call them gap formations in the direction of the fibers rather than the concentric formation of gaps. It is difficult to understand why in some cases, in otherwise clear lenses, parallel lines of separation extend across the lens (from one edge of the pupil to the other) without regard to the suture system (Fig. 396 A, B). In these cases there is no interruption or change in direction as they pass

over the sutures. While these lines usually seem to extend between the radial sutures, at times they are interrupted and appear like the cross threads of a spider's web (Fig. 397). The visibility of these

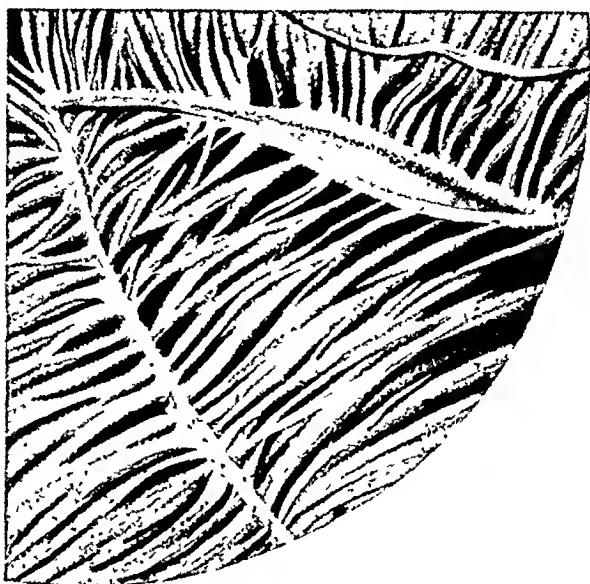


FIG. 397. Lamellary separation. Separated lamellae between sutures resemble the crossing fibers of a spider's web. (Plastic model after Vogt.)

structures varies with their direction and the angle between observation and illumination; like the radial water slits they are best seen when the zones of specular reflection are approached. By slowly changing the angle of illumination the lines appear more or less distinct. In addition if the direction of the lines suddenly changes or differs from that seen with one position of the light, a change in the angle of illumination is necessary in order again to make them visible. In cataracta cuneiformis, white concentric lines of reflection are found in the region of the flat opacities themselves or just axially to them (Fig. 395). They run in a direction that is at right angles to the apices of the spearlike cuneiform opacities and may be located in front of or behind them.

In addition to those that extend frontally in apparently a single layer, the optic section frequently reveals the sagittal extension of small white lines through the cortex. Here, it will be seen that groups of short whitish parallel lines separated by dark intervening spaces traverse the cortex in a horizontal or oblique manner (Fig. 398).

The exact explanation of these sagittally directed lines, which commonly cross concentrically directed water slits is not known. They may represent the opacification of radial lamellae which are sectioned

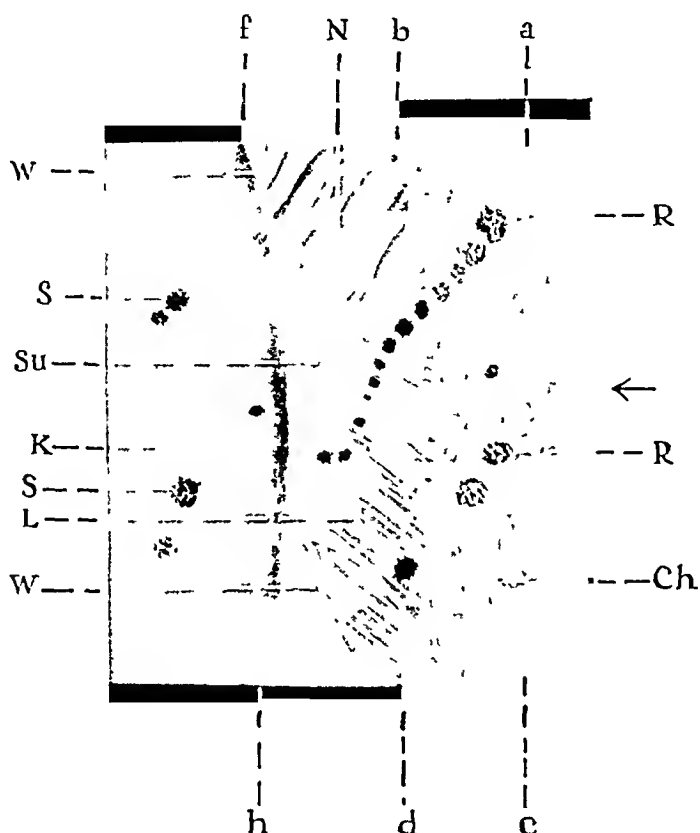


FIG. 398. Cortical cataract (incipient) as seen by direct focal illumination (wide beam). Pigment on the anterior lens capsule casts shadows. Water slits (dilated suture lines at the level of the surface of the adult nucleus). Note obliquely running white lines of lamellary separation. *a, b, c, d*, surface of the lens (broad beam [parallelepiped]); *b, d, f, h*, surface of adult nucleus; *R*, iris pigment deposits; *Ch*, shagreen of anterior lens capsule; *f-h*, deeper edge of parallelepiped or surface of adult nucleus; *K*, opaque adult nucleus; *Su*, dark suture on the surface of opaque adult nucleus; *L*, white lines (lamellary separation) located between capsule and nucleus; *W*, border of water cleft; *S*, dark spots (to the left) representing shadows cast from the pigment deposits on the anterior lens capsule when viewed by light (retro-illumination) from the deeper parts of the lens; *N*, surface of the adult nucleus. (After Vogt.)

by the narrow beam in their radial extension, in other words, lamellary separation of the radial lamellae. The dark gaps between the white lines may represent either the less relucant fluid which has separated and compressed lamellae, or groups of fibers similar to that which occurs in sutures in the formation of radial water slits. The direction of these lines like the longer ones, seen frontally and described above, in most instances tends to run obliquely from tem-

porally below to nasally upward. In one of Vogt's cases the white lines crossing a frontally directed water slit had a flared (?) and wavy appearance similar to those seen in diffuse illumination of the

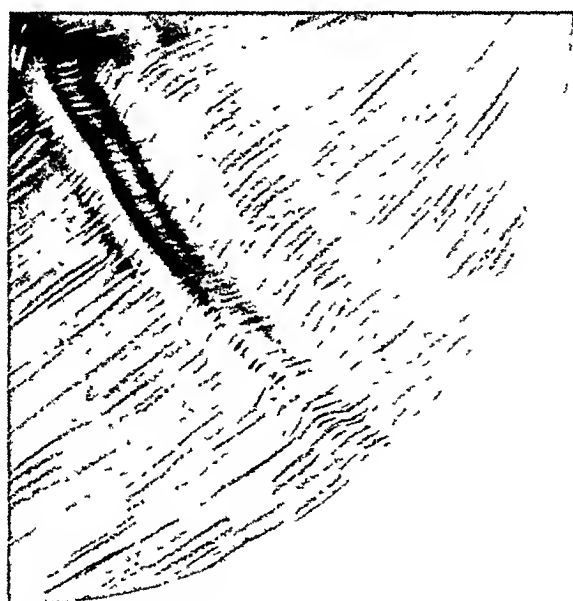


FIG 399. Lamellary separation showing the bending of the lamellae as an expression of the shrinking of the nucleus in the aged.

adult nucleus relief. He explains the bending of the lamellae as an expression of shrinking of the nucleus in the aged (Fig. 399). Owing to the diffuse reflection from the anterior parts of the lens, lamellary separation of the posterior cortex is difficult to see. Vogt states that it does occur and when present the direction of the lines is predominantly opposite to the anterior cortex, namely from below nasally to above temporally.

The optic section often reveals that these whitish sagittally directed lines of separation are related to flat and frontally directed concentric water slits. It has been shown that in many instances this type of water slit is nothing more than the axial extension of the flat cuneiform opacities. The flattened cuneiform opacities are formed by myelin droplets in the walls of concentric water slits, in the same way that spokes develop from radial water slits. The sagittally directed lines mentioned above do not necessarily have to cross the entire cortical thickness but being short may only just extend across the front or back wall of the concentric slit. At times, particularly

in intumescent cataract, small groups of vertically parallel white lines (lamellary separation) may be seen in various parts of the cortex without any apparent connection with the frontally directed water slits. However, it should be mentioned that these slits are often very difficult to see, requiring not only the employment of the narrow beam but constant readjustment of the angle between illumination and observation.

Schild in Vogt's Institute⁵⁹⁹ examined the eyes of 218 old persons. Lamellary separation was not found before the fiftieth year (although Vogt himself saw it in younger persons). He found evidences of it in 7 per cent of persons between 50 and 60 years; 18 per cent between 60 and 70 years; 52 per cent between 70 and 80 years and in 50 per cent over 80 years of age. The direction of the lines of laminary separation was predominantly from temporally below to nasally upward. The spider-web form was infrequent (Fig. 397). In 75 per cent it was found to be bilateral and apparently was rarer in the posterior cortex.

CUNEIFORM CATARACT (CATARACTA CUNEIFORMIS PLANA PERIPHERICA)

Cuneiform opacities constitute the most common form of cortical alteration confined to the periphery and as such play an integral part in the formation of senile cortical cataract. Characteristically they form flattened opacities (sagittally thin), which like coronary cataracts may extend posteriorly around the equator to involve the posterior cortex (Fig. 400). Their peripheral edge usually runs parallel to the equator while their axially directed ends form broadly rounded, pointed or dentate processes. Although located in about the same depth of the cortex as coronary opacities, the flat pointed, angulated cuneiform opacities differ in appearance from the more rounded discrete clublike processes of coronary cataract (Plate LXVII, fig. 6). It should be borne in mind also that coronary cataracts develop at an earlier age. Cuneiform opacities, like the lines of lamellary separation with which they are usually associated, are

localized mostly in the lower nasal periphery, another difference from the coronary opacities, which are found circumferentially around the equator of the adult nucleus. Fusion of the axial dentate



FIG. 400. Unusual type of cuneiform opacities. (After Vogt.)

cuneiform extensions may result in broader segmental opacities (Fig. 395).

Frequently concentric stripings are seen running at right angle just axially to the flattened cuneiform opacities. The innermost part of these concentric lines often blend into laminary separations. Cuneiform opacities, like most of the other presenile or incipient cortical changes, tend to progress with time but with the usual unpredictable and individual variability. Axial progress is particularly noticeable posteriorly.

The concentric stripings may also run directly over the opacities themselves and probably indicate the close relationship between cuneiform cataract and the concentric lines of separation. Not infrequently cuneiform opacities are combined with flat (sagittally directed) water slits and radial spokes. The optic section will demonstrate the relative thinness of the opacities but despite this they

appear white in color by focal light. Retro-illumination especially of those in the posterior cortex will reveal a yellowish to brownish tinge (because of the yellowish color of the deeper parts of the lens in age). According to Vogt, the frontally flattened direction of cuneiform opacities and their relation to laminary separation supports the conception that the lens is composed of concentric (onion-like) lamellae. He holds that cuneiform cataract develops from the frontally flattened concentric water slits formed by the accumulation of fluid between the concentric lamellae. Opacification of this type of water slit (similar to the radial slits where opaque spokes are formed) results in cuneiform opacities. Moreover, as long as the slit is not opaque, we get the picture of laminary separation, a consequence of the separation of fiber lamellae by fluid. Cuneiform opacities are commonly bilateral and, similar to other presenile changes, may be genetically determined.

PRESENILE AND SENILE PUNCTATE OPACITIES (CORTICAL)

Biomicroscopically, it is rare to find an adult lens without an occasional small, white, punctate dot in the cortex. They are so delicate that they cannot be observed with the loupe or with the ophthalmoscope. As such, they have been considered as physiologic but since they tend to increase with age and at times even interfere with vision, they have been properly classified under presenile lens opacities. These dots or small lines are especially prevalent in peripheral coronary cataracts. With advancing age they increase in number, especially in the equatorial region of the cortex (Fig. 368). They are seldom found in the axial regions. At times in the aged they may fill the peripheral cortex appearing as dense snowflake opacities or as a fine dust. Occasionally, instead of being isolated or disseminated irregularly through the cortex, they may develop into linear stripes, deep in the equatorial region of the cortex, and following the nuclear curve, form hooklets not unlike those seen in zonular and coronary cataract. As a matter of fact cases have been seen in which coronary cataract apparently developed from, or at least was

preceded by, the presence of fine punctate dots peripherally. Small dot- or dustlike opacities are often associated with the various forms of senile cataract (cortical, nuclear, or cupuliform) as well.



FIG. 401. Concentric lamellar opacities (cuneiform type) extending around the adult nucleus equator. Optic section.

SENILE PERIPHERAL CONCENTRIC LAMELLAR OPACITIES (VOGT)

When the diffuse dustlike opacities mentioned above, increase in number toward the equator, they may become condensed into concentric lamellae. They may form several layers separated by clear lens substance and when in the deeper cortex, extend parallel to the direction of the adult nuclear stripe (Fig. 401). The deeper ones may outline the nuclear equator as they round it to reach the corresponding layer posteriorly. It is necessary to employ the narrow beam in order to see the successive layers clearly. Since these opacities seldom reach the axial regions, they do not affect vision and are easily overlooked unless a mydriatic is used. Because of their location they may be confused with coronary opacities. In the early stages, with the pupil well dilated attention may first be called to such a change by the increased relucency of the adult nucleus near the equator in connection with the presence of small white dots. Concentric peripheral lamellar opacifications are usually found in association with other senile changes but not necessarily so. It has been found that the opacities vary from 0.8 to 1.2 mm. in width.

AXIAL PUNCTATE OPACITIES IN THE ANTERIOR CORTICAL
SUTURE SYSTEM

This is a characteristic type of opacity which, with lower powers of magnification, seems like a hazy delineation of the cortical sutures



FIG. 402. Axial punctate opacities in anterior cortical suture system.

(Fig. 402). With higher power it will be seen that the haze is composed of minute dots, varying in color from gray to yellow, brown, or even reddish or coppery hues.* It is obvious that opacities of this kind, which are just barely visible with the biomicroscope, do not disturb vision in themselves; but since they occur in the older age groups, it is to be expected that they will be associated with the other changes of senescence. The latter (cortical and nuclear opacifications) which increase reflection posteriorly may add further difficulties in detection of the delicate punctate cortical suture opacities unless the narrow beam (optic section) and careful focusing are employed.

* Although apparently occupying the cortical sutures, this type of opacity is a stationary or very slowly advancing process. Vogt first saw such lesions in 1919 and states that probably because of their delicacy they were overlooked before this time. They are not connected with water slits or spoke formation because (1) there are no water slits in the vicinity of the affected sutures, (2) the microscopic dots are much smaller than any of those seen in water slits or spokes, and (3) the dots are of regular size with no tendency to coalesce into vacuoles. In addition, at times the tiny dots are colored and glitter like crystals. That this type of opacity is chiefly found in adults and the aged was shown by the compilations of Müller and Rehsteiner⁵⁵⁰ who examined 267 institutional inmates. They found punctate anterior cortical suture opacities in 22 cases, or 12.1 per cent. The ages varied from 40 to 90 years. In a group of 19 persons, aged 40 to 50 years, they found one case; while in a group of 70 persons, aged 61 to 80 years, they found 16 cases. Vogt's youngest case was aged 36 years.

Starting in the axial region, the figure follows the design of the cortical suture system and branches out in radiating directions, gradually fading out in the outer one-third. Mostly the figure lies subcapsularly in the anterior parts of the cortex and only in one case did Vogt find it deeper, near the adult nucleus. He found that their length varies from 0.5 to 2 mm. and their width from 0.05 to 0.2 mm. However, all the suture branches of the system may not be affected. Sometimes the dots appear like glittering points. The reason for these color variations has not been explained, but it is similar to the hypothesis advanced for the color differences of cerulean opacities. Occasionally only a small group of dots (abortive form) may be present subcapsularly in the axial area of the suture or the point of fusion of the converging branches. In this case, of course, the radiating opacity will not have any branchings.

IRREGULAR CREVICES OR FISSURES IN THE ANTERIOR CORTEX IN MATURE AND HYPERMATURE CATARACT (VOGT)

Vogt described the finding of dark, irregular, more or less radial fissures in the anterior cortex of mature cataracts. They stand out sharply as dark cracks within the opaque cortical mass by contrast. At times the sharp edges of the fissures have a dentate appearance and, like a gear, appear to mesh with those of the opposite side (see Figs. 403 and 405A). These sharp-edged fissures differ from water slits in that they extend through the cortical thickness and probably result from shrinkage rather than from distention. Vogt stated that they reminded him of fissures formed in dry earth. Their radial direction may signify a relationship to the suture system. The width of the fissures vary from place to place. Frequently they have smaller terminal branches. In direct focal light, the passage of the beam into the depths may reflect a reddish color from the nucleus not unlike that of cataracta brunescens or nigra. Vogt also described similar dentate radial fissures within isolated lancet-like spokes of the anterior cortex. His impression was that these fissures originate in the cortex by retraction of its opaque substance and that it is not

dependent on nuclear shrinkage *per se*. In any event, these ruptures are a late manifestation and differ in their genesis and time of appearance from the radial water slits, although the fissures may occur

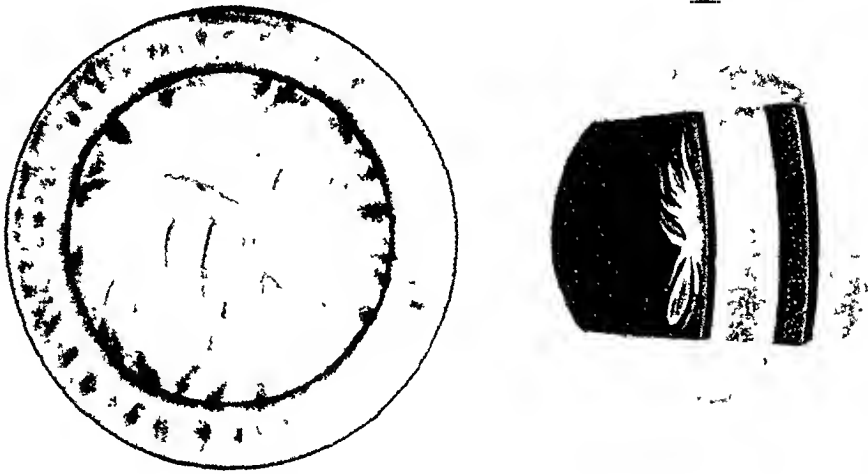


FIG. 403. Intumescent to mature cataract after paradichlorbenzine poisoning.

within them. Similar crevice-like formations have also been seen in total traumatic cataract.

IMMATURE CORTICAL CATARACT (INTUMESCENT CATARACT)

As a result of the progressive development of water slits, spokes, lamellary separation and cuneiform opacities, punctate dotlike opacities, etc. — all indications of hydration and subsequent dissolution of the lens fibers — the lens swells. Clinically the process of swelling may make itself manifest by a decrease in depth of the anterior chamber. The degree and rapidity of swelling or intumescence varies greatly depending on the intensity of the causative factors and the reaction of the lens to them. There are cases in which the stage of intumescence is short-lived, the cortex rapidly becoming completely opaque and the cataract mature. This is especially true in some types of toxic and traumatic cataract. In a case that I reported (paradichlorbenzine cataract) ³⁶⁷ the changes proceeded with such rapidity that within a week the vision deteriorated from normal to the bare recognition of hand movements. Owing to the rapid swelling of the lens, which became entirely opaque in a few days, the

intra-ocular pressure suddenly rose to such a high degree that on the seventh day following the inception of the cataractous changes, a linear extraction had to be performed (Fig. 403). The presence of sclerosis (nuclear) which may also effect the cortical fibers to a lesser degree, may act as a deterrent to intumescence. However it is not surprising that in younger persons, rapid swelling can occur in both the cortex and nucleus (Fig. 405 B). Although ordinarily the stage of intumescence is marked by an increase in water content as seen by the presence of numerous water slits and zones of lamellar separation in the beginning, opacification may be comparatively moderate, consisting of radial spokes and peripheral cuneiform opacities and dots, more or less limited to the deeper cortex (Fig. 398, p. 1119). As these changes progress, the cortex has a hazy appearance, when seen by diffuse light. Grayish broad bands of opacity may converge from the periphery to the axial regions. The increase in whiteness of these spokes indicates their older age. These radiating band opacities may form rosette figures. At times the opaque spokes become fibrillated so that instead of being uniformly opaque, they appear to be composed of irregular branching radial white lines. Between these lines roundish opaque spots (opacified vacuoles) may be seen. Or the spokes, especially toward the periphery, may appear as homogeneous white lanceolate areas. In front of these dark or opaque spokes small subcapsular spots indicate the products of vacuolar degeneration.

Optic section will reveal large, often gaping, water slits surrounded by grayish zones (Fig. 404). Groups of more or less parallel whitish lines running obliquely will be found in various places within the cortex. This type of laminary separation indicates that the fluid, which in the early stages resulted in the formation of clear water slits (radial and frontal), now has separated individual lens fibers and has caused them to become opaque (Fig. 398; Plate LXVII, fig. 7). It frequently happens that through manipulation (massage of the capsule with the iris repositor) either purposely or accidentally during the performance of a preliminary iridectomy in cases in which nuclear cataract and only minimal cortical changes

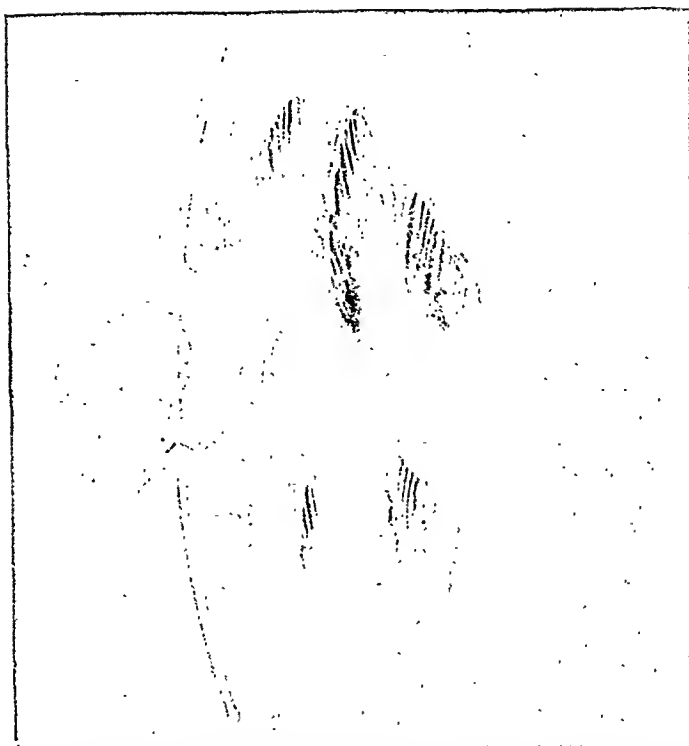


FIG. 404. Intumescent cataract. Cortex shows large water slits. Note central disc-like capsular opacity.



A



B

FIG. 405. A. Intumescent to mature cataract. Note clearer subcapsular layer composed of vacuolar material. Viewed in optic section. B. Vacuolar type of cortical and nuclear cataract seen in a young person.

exist, that a sudden intumescence results within a short time following the operation. The progression of the intumescent stages can then be easily observed — especially the formation of the broad opaque bands (opaque spokes) and of individual fiber opacification suggestive of the appearance of experimental maceration *in vitro*. Another interesting finding, even in the advanced stages of intumescent cataract, is the occasional presence of a relatively dark (clear) space between the main mass of cataractous cortex and the capsule (Fig. 405 A, B). In other words except for the tendency to vacuolar formation, the immediate subcapsular zone, which consists of the very younger fibers, tends to remain clear for a long time. This clear fluid-like space may be continuous with deeper radial water slits. At times in optic section near the main deeper mass of cortical opacities, one or more thin opaque layers of opacity may be found within the clearer subcapsular fluid layer. Vogt believes that such an independent layer represents the more solid continuity of fibers of a zone of discontinuity as compared to the intervening areas. The increase of curvature or bulging of the capsule can frequently be seen with the optic section. This method also reveals an actual increase of thickness of the cortex (Fig. 405 B). This can be verified by the fact that the anterior zone of specular reflection is smaller at any one given point than that found in a lens of normal capsular curvature. Because of an increase in curvature the number of specularly reflected rays from any one point will of necessity be smaller. This is exemplified in the normal lens in which, owing to the greater curvature of the posterior capsule, the shagreen area is always smaller than that of the flatter anterior capsule. Examination with focal light often reveals the association of pre-existing cuneiform and coronary opacities in the cortical periphery. However, it is still questionable whether the presence of coronary opacities influences or predisposes to the formation of the changes typical of senile cataract. I have in mind one case (Plate LXVI, fig. 5), that of a 60-year-old woman under my observation for at least fifteen years, who had especially marked bilateral cerulean and coronary opacities. At the present time her central vision is still 20/20 in each eye.

There are no other cortical presenile changes present but, as is to be expected, there is a slight increase in nuclear relucency. That similar changes to those of the anterior cortex occur in the posterior cortex during intumescence is undoubted. However, owing to accompanying anterior cortical changes, the inevitable nuclear cataract often present, and posterior saucer-like opacities (all resulting in increased internal reflections) examination of the posterior cortex becomes difficult during the intumescent, mature, and hypermature stages, but it is surprising how often examination of the deeper layers is feasible despite these changes.

RARER FORMS OF SENILE LENS CHANGES

Vogt has drawn attention to rarer forms of cortical senile cataractous changes that do not follow the ordinary patterns. Among these are the ring forms and disciform opacities found primarily in the axial regions. The former, found in the deeper cortex, consist of angulated opaque bands roughly forming a ring. Within and mixed with it there may be intensely white spots. The ring may be formed by opacification of the edges of the radial water slits in the axial region. Similar ring forms in the anterior cortex may occur in complicated cataract such as in myopia and glaucoma. The disk form is usually located in the deeper cortex. It may appear as a thin circumscribed opacity, composed of fine dots. In the case described by Vogt, the anterior surface of the disk was flat and the posterior surface was convex.

MATURE CATARACT

From the morphologic standpoint a cataract is considered to be mature when the opacification and disintegration of the cortex becomes complete, i.e., when the opacification actually or almost, reaches the capsule (Fig. 405 A). Unless the central part of the lens has been involved by other types of opacities, vision may still be serviceable during the incipient and early intumescent stages of the cataract. But as maturity approaches vision becomes markedly reduced to a degree that the patient can barely count fingers or detect

hand movements. Finally only the ability to perceive light is retained. The stage of maturity is marked by dehydration and by an increase in coagulation of the cortical substance. This loss of water is evidenced biomicroscopically by a reduction in swelling (flattening of the lens with increase of chamber depth) and by decrease in the size and number of the clear dark water slits. The chief feature is now an increase in opacification. The radiating bands (spokes) mentioned above become more intensely white, and, instead of being separated by clear fluid, it will be seen that they are crossed by opaque fibers or round or irregular opaque masses. The radial spokes themselves may become wavy or even broken up into smaller lanceolate opacities, so that very little, if any, indication of the original spokelike appearance remains. Larger, coarse vacuoles are not unusual (Fig. 405 A, B). In direct focal light these may appear as dark or black holes within the opaque cortical substance, whereas their true vacuolar nature can be demonstrated by indirect or retro-illumination. Carpets of subcapsular or perhaps epithelial vacuoles in direct focal light appear as delicate thin whitish opacities. Likewise with high power and indirect or retro-illumination it will be seen that these are composed of groups of very fine vacuoles. Vogt has described in great detail the various ways in which opaque fibers and darker lines may traverse spokes. In some cases delicate, more or less parallel white lines may cross a spoke at right angles to its radial direction. In others, dark lines corresponding to the dark spaces between individual opaque fibers may run radially in an irregular way, sometimes forming sinuous figures. He thinks that the former may represent a cross section of concentric lamellae, and that the latter represent changes in the radial lamellae. There is no reason to doubt that changes of maturity, similar to those that occur anteriorly, also occur in the posterior cortex. But, as pointed out in the section on intumescent cataracts, the latter are difficult to see except on the rare occasions when they have developed before nuclear and anterior cortical changes have precluded a clear view of this area.

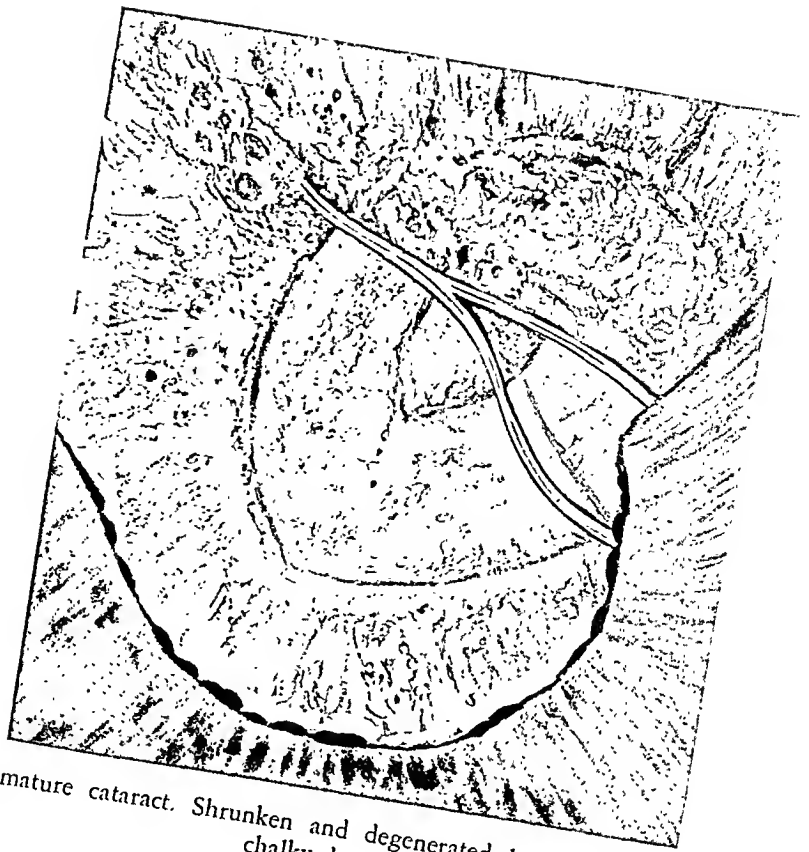


FIG. 406. Hypermature cataract. Shrunken and degenerated lens. Note capsular folds and chalky deposits.

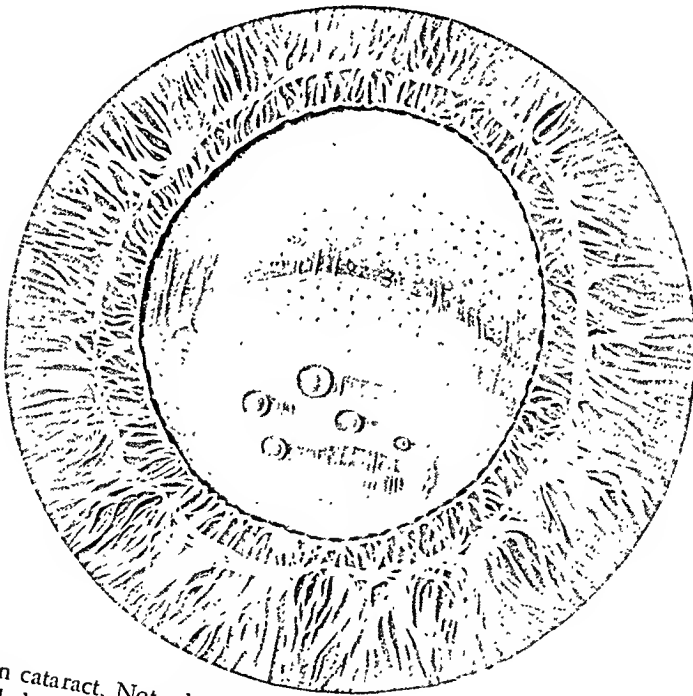


FIG. 407. Morgagnian cataract. Note downward displacement of the nucleus; vacuoles; striated folds and small dots (colored cholesterol crystals). (After Duverger and Velter.)

HYPERMATURITY CATARACT (CATARACTA HYPERMATURA)

Hypermaturation of cataract in a gross way might be compared to the healing processes observed in other tissues. In the lens it is characterized by elimination of the degenerated lens products, in so far as diffusion of these substances via the lens capsule is possible, and by subsequent contraction and secondary organization of the cortical remains (Fig. 406). The latter is illustrated by the frequency of calcareous deposits and crystals. Eventually this process leads to great shrinkage so that one may see a wrinkled capsule (at times unevenly curved owing to small concave depressions)* containing a yellow nucleus, which is surrounded by degenerated cortical material. Consequently, there is a deepening of the anterior chamber. Just below the wrinkled capsule, numerous vacuoles may be found by indirect illumination (Fig. 407). The nucleus may be displaced and, owing to degeneration of the zonule, the shrunken lens may become dislocated. In such cases iridonesis and phakodonesis may result. In extreme cases the lens may be converted into a thickened membrane or even, as has not infrequently been reported, a "spontaneous cure" results since retraction of this membrane by the zonular filaments produces a veritable aphakia.

From the standpoint of biomicroscopy the chief initial indication of hypermaturation is the presence of capsular folds. The subcapsular cortex usually has a milky white appearance (beginning liquefaction), being composed of white clumped granular opacities. In a case recently observed, there were fairly small white flakelike particles (bilaterally) in the anterior chamber. At first it was thought that these were indications of an inflammatory process. There were no keratic precipitates. The irides were normal and the eyes were otherwise quiet. There was no peeling of the capsular lamellae visible, but one or two capsular folds were found. So far as could be seen no rupture in the capsules were noted. These flakes were observed unchanged for some months until the cataracts were operated on.

* Apparent bending of the anterior capsule may result from an optical artefact following irregular astigmatism caused by corneal scars. When considering irregularities in capsular curvature one should not forget to eliminate corneal scars as a factor.

The recovery was prompt and eventful. . . . Evidently these particles were degenerated lens particles (capsular or cortical?). The presence of such broken-down lens particles in the aqueous could in sensitized individuals set up an anaphylactic reaction and a toxic iritis.

Crystals. The finding of glittering crystals is not unusual in mature and hypermature senile cataracts, as well as in complicated and traumatic cataracts. They may occur as colored shining points or as long needles, presumably cholesterol (Plate LXVIII). Such crystals can be found at times, especially in older persons in a relatively clear sclerotic lens. But they also have been seen occasionally in otherwise normal lenses of children as a congenital condition. The direction of the needles may follow that of the radiating lamellae (especially at the periphery) where they may occupy spaces within the opaque cortex that correspond to the suture. In the axial region, however, groups of long varicolored glittering needles may traverse the deeper cortex and nucleus, apparently following no orderly pattern. At other times they may form leaves or layers. Smaller ones may be found (when lens transparency permits) near the embryonal Y-suture. Of themselves, like vitreous crystals, they do not interfere with vision.

Capsular Opacities. Another sign of maturity and hypermaturity, well known for a long time, is the presence of *small capsular opacities*. These may be the result of the opacification of hypertrophied capsular epithelium (Fig. 404). The spots in diffuse or focal light are grayish to white in color and generally are more or less circular. In one case the measurements of a comparatively large one was horizontally 0.56 mm. and vertically 0.4 mm. Like other capsular defects, which have a sudden level change at their borders, when observed in specular reflex these have a "shagreen free halo" (page 974). When the opacity is observed in the mirror reflex, bright linear stripings will be seen frequently. Vogt has interpreted these as fiber formations brought about by epithelial proliferation. At times these stripes may glitter like crystals.

Complete liquefaction of the cortex results in the so-called "morgagnian cataract" (Fig. 407). In these cases, which are compara-

PLATE LXVIII

FIG. 1. Crystalline (cholesterol) degeneration within the adult nucleus.

FIG. 2. Layer of fine crystalline deposits beneath the anterior lens capsule, corresponding to the anterior line of disjunction.

FIG. 3. Crystalline deposits in total cataract.

FIG. 4. High-power view showing the details of crystals seen in case in Figure 3.



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tively rare, the yellow sclerosed and opaque nucleus may be found almost floating within a fluid milky cortex or, owing to the effects of gravity, may sink somewhat within the saclike capsule. Vogt described three cases in elderly men having mature to hypermature cataracts in whom a sudden iritis with increased intraocular pressure appeared. The toxic iritis apparently results from the presence of cortical material in the anterior chamber. In all three cases the iritis and secondary glaucoma subsided after iridectomy, and was followed by a rapid shrinkage of the lens. The opaque nucleus sank, leaving a clear intact capsular sac above containing faintly relucet fluid with a few crumb-like pieces of opaque lens matter adherent to its inner walls. This kind of cortical shrinkage differs from that seen in hypermature cataract characterized by folds of the capsule, in that in the latter the capsule remains in close contact with the cortical remains and shrinks with it. In the former the capsule is separated from the shrunken cataractous mass and extends either tautly or relaxed in the direction of the zonular attachment. I recently saw a case in a 70-year-old man in whom the left eye had been operated on several years previously for removal of a cataract. There was a shrunken lens (hypermature) in the right eye. He suddenly developed pain and congestion in this eye, the pressure rising to 60 mm. (Schiötz). After an iridectomy, the eye quieted down, and it was possible to see a clear capsular "bag" in the coloboma. Some weeks later the shrunken lens was extracted intracapsularly with no complications, and eventually 20/40 vision was obtained. In the free part of the anterior capsule the shagreen was still present. This might strengthen the contention of Butler, that the shagreen is the function of the hyaline capsule itself. In this case any reflection from the epithelium and superficial lens fibers (Gullstrand and Vogt) might only be auxiliary to the formation of the shagreen, in which no epithelium is present. In Vogt's cases the anterior capsule in the free part seemed slightly more taut than the posterior. Also both parts became flaccid following the instillation of pilocarpine drops.

Capsular Folds. The presence of capsular folds is always an indication of shrinking of the lens cortex. Before the days of bio-

microscopy, clinical descriptions of hypermature cataracts included little concerning the presence of capsular folds. Because of the underlying opacification, it is ordinarily impossible to determine

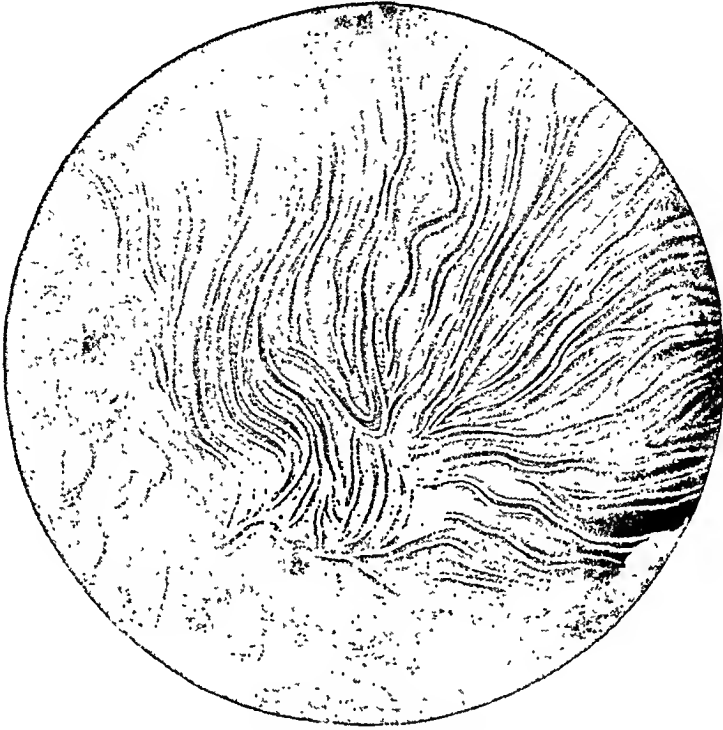


FIG. 408. Capsular folds.

whether the posterior capsule undergoes folding, although anatomically this is quite possible. Folds of the anterior capsule occur not only in hypermature cataract but also in cataracta complicata and in traumatic cataract as well. In complicated cataract it has been noticed that dispersed iris pigment can be deposited on folds in the form of brown stripes. Vogt mentions the presence of capsular folds in cases of so-called "soft juvenile complicated cataract" where they have bluish tints in direct focal light. Ordinary folds, like vacuoles, are best seen as doubly-reflecting lines in retro-illumination or by indirect illumination (Fig. 406). With this form of illumination they remind us of folds of Descemet's membrane, frequently having an irregular "wormlike" course and often branched (Fig. 408). Their average width has been estimated as from 0.05 mm. to 0.5 mm. However, like folds of Descemet's membrane, their length varies greatly, and it is not unusual to follow a single fold across the

entire pupillary opening. Often, owing to the opaque background of the cortex, these capsular lines cannot be seen distinctly in direct focal illumination.

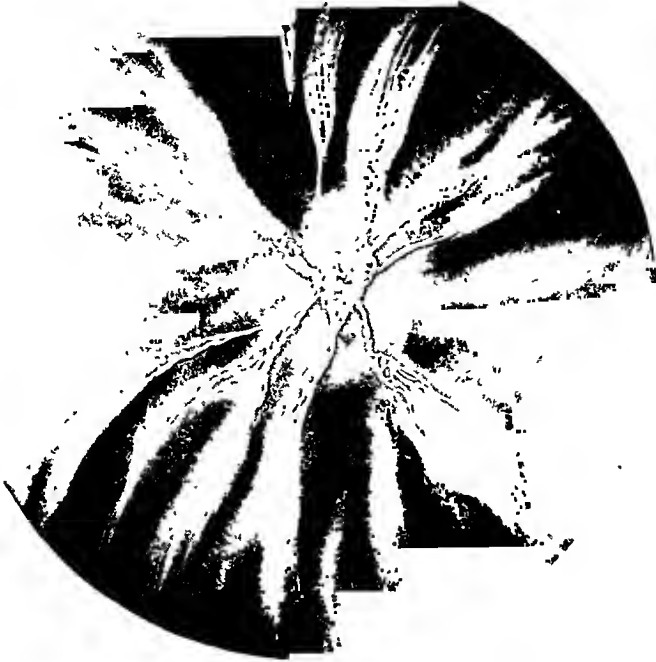


FIG. 409. Anterior and posterior saucer cataract. Diffuse view.

CUPULIFORM CATARACT (ANTERIOR AND POSTERIOR SAUCER-SHAPED CATARACT — CATARACTA SCUTELLARIS)

Although the so-called "saucer-shaped" cataract is commonly considered as principally posterior (just in front of the posterior capsule), a similar type may be found just below the anterior capsule (Fig. 409). Characteristically, these forms of senile opacity are usually restricted to a single layer and are composed of vacuoles and granular material. In most instances they tend to be more densely developed in the axial regions and hence cause visual disturbances at an early stage.

Anterior Saucer Cataract. Anterior saucer cataract is usually associated with posterior saucer cataract or with other cortical and nuclear senile lens opacities. In diffuse illumination it appears as a whitish starlike figure, the branches of which radiate from a central

PLATE LXIX

FIG. 1. Anterior cupuliform (saucer type of senile cataract) cataract. Diffuse illumination.

FIG. 2. Same as case in Figure 1, as seen by direct focal illumination. Optic section.

FIG. 3. Posterior cupuliform (saucer type). Early. Diffuse illumination.

FIG. 4. Same as case in Figure 3, by direct focal illumination. Optic section.

FIG. 5. Anterior and posterior cupuliform (saucer-like) opacities. Composite view. Direct focal illumination. Optic section.

FIG. 6. More advanced posterior cupuliform (saucer-like) cataract. Diffuse illumination.

FIG. 7. Same case as shown in Figure 6, showing details of vacuolar degeneration. Direct focal illumination. High power.

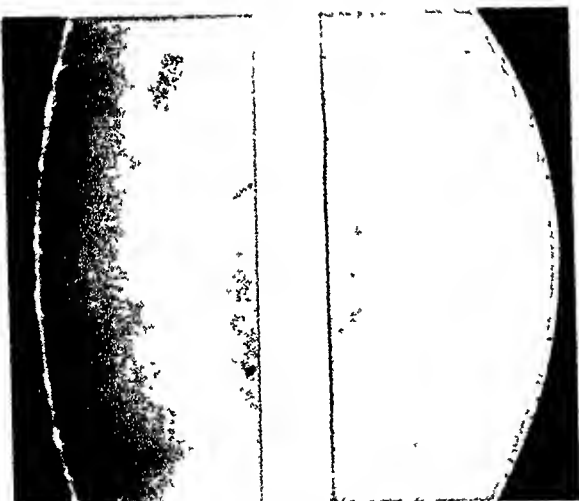
FIG. 8. Same as case shown in Figure 7, viewed in specular reflection.



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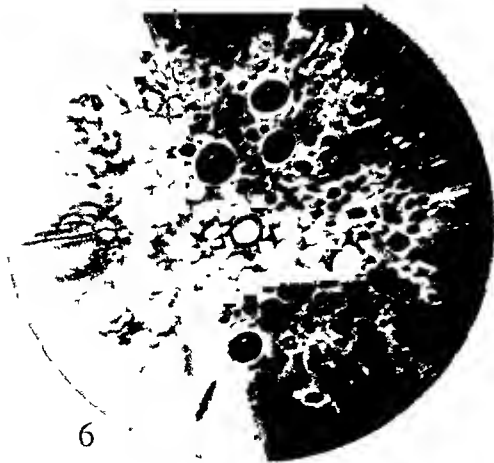


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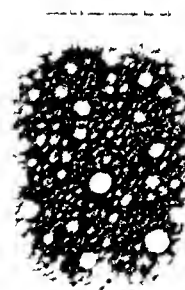
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axial opacity. (See Fig. 406; Plate LXIX, figs. 1, 5.) However, in the early stages it may not show any radiations and resembles the faint subcapsular opacities observed in mature and hypermature cataracts which by retro-illumination are seen to be composed of fine vacuoles. Located subcapsularly, the opacity tends to be thin and lies in one plane. In direct focal light the grayish opacity appears sievelike with dark round holes, reminiscent of the structures seen in band-form keratitis. (See Vol. I, page 345.) These dark holes represent vacuoles the contents of which are less relucant than the surrounding gray area. The lesion starts in the axial region and progresses very slowly in a radiating manner.

Posterior Saucer Cataract. Posterior saucer cataract is one of the common forms of senile cataract. It is characterized by the presence of a single (rarely double) layer of opacity adjacent to, or just in front of, the posterior capsule (Plate LXIX, figs. 5, 6, 7, 8). Similar to the anterior variety, it is composed of a thin sievelike vacuolar and granular substance. Before the days of biomicroscopy, its exact location, i.e., in front of the posterior capsule, was not recognized. As seen with the ophthalmoscope (when well developed) it was considered as a posterior lamellar cataract. When combined with nuclear cataract or fairly well-developed cortical changes, the ophthalmoscopic diagnosis of posterior saucer cataract may become difficult. Because of its axial location, visual loss is rapid and extraction may be required in spite of the black appearance of the pupil. In this way it differs from the other types of senile cataract, e.g., incipient cortical and nuclear opacities, which may require years or decades before seriously interfering with central vision. It is rare indeed to find posterior saucer cataract developed to any degree without the concomitant presence of nuclear cataract and at least a few signs of incipient cortical changes (water-slits, spokes, cuneiform opacities, etc.). When the opacity is centrally located, it may be seen through an undilated pupil, provided the angle between illumination and observation is very narrow. However, frequently the opacity may be paracentral or peripheral even in its early stages and then dilation of the pupil becomes diagnostically necessary. With the narrow

beam its exact situation is easily determined as well as its peculiar composition. When employing the prefocal or postfocal part of the beam (diffuse light), its entire saucer-like shape, corresponding to

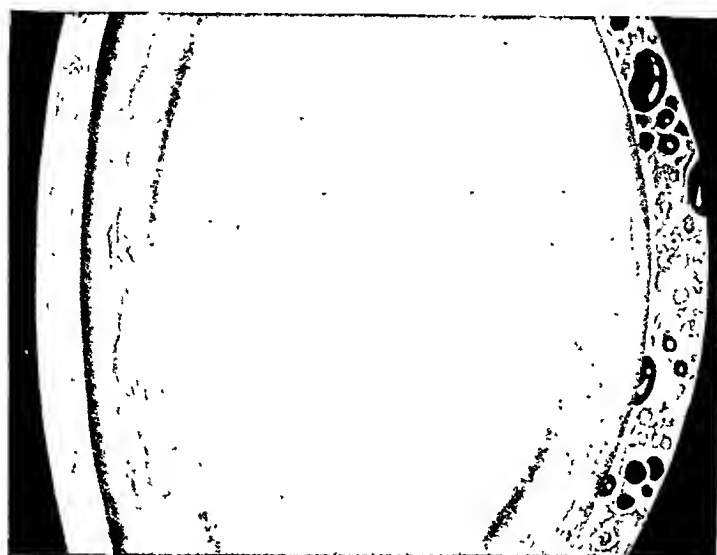


FIG. 410. Posterior saucer cataract with a few anterior subcapsular opacities arranged in a star form.

the curve of the posterior capsule, and its composition can be seen. It has been pointed out that the appearance of the opacities varies although their different aspects may blend one into the other. First there is the type in which the opacity appears as a thick yellow mat with vacuoles within or superimposed on it. The vacuoles in this form resemble small circles or rings with dark contours and light yellow centers. The second form is thinner (allowing transillumination with the light of the ophthalmoscope) and resembles a sieve. It consists of a thin grayish background punctuated by dark round holes of varying sizes and in general not unlike the appearance of the opacity of band-form keratitis (Fig. 410). (See Vol. I, page 345.) As the substrate of the latter type thickens, it tends to form an intermediary type approaching the thicker and yellower first form. Occasionally a posterior saucer cataract may be composed of a double layer, the ends of the anterior one joining the one behind it peripherally, resulting in a sort of meniscoid structure. The tendency to the formation of a similar meniscus-like double layer opacity has also been reported in irradiation cataract.

It is interesting to speculate why this type of change, which in some ways resembles *cataracta complicata* so that at times it is difficult to differentiate between them, is so characteristic of the deeper parts of the posterior cortex. Perhaps the absence of epithelium is a factor. Also it may be that the process of nuclear sclerosis extends backward into the posterior cortex to a greater extent than it ordinarily does anteriorly, leaving vulnerable only a thin sub-capsular layer of young fibers. At any rate posterior saucer-shaped cataracts spread out in a flat way in front of the posterior capsule and follow its curvature. Its anterior surface is more or less sharply defined showing little or no tendency to invade the posterior cortex or posterior adult nucleus in an anterior direction. This, together with the more porous consistency and the vivid color display in complicated cataract, helps to differentiate them.

NUCLEAR CATARACT

Undoubtedly the biomicroscopic examination of the lens in older persons has demonstrated the high frequency with which the opacification of the nucleus occurs and the important role that it plays in contribution to senile cataract. We are now able not only to discern the earliest manifestation of nuclear change but also to follow its customary slow development over long periods. With the narrow beam, its presence and progress can be determined even when cortical changes are marked. The characteristic change of nuclear cataract, i.e., a uniform cloudiness, does not cause severe visual impairment (as compared to cortical or cupuliform cataract) until discoloration leads to *cataracta brunescens* or *nigra* or is complicated by the formation of posterior saucer cataract. In my opinion, it is very rare indeed not to find posterior saucer cataract and anterior cortical changes associated with a well-developed nuclear cataract. This might indicate that nuclear cataract may play a role in the formation of cortical changes. On the other hand, the nucleus often may remain relatively clear for a long time in the presence of an advancing cortical cataract, but in the end it always participates in the process of opacification. It should be pointed out that nuclear opaci-

fication is not only a symptom of senile cataract but may also accompany rapidly developing cataracta complicata, especially those secondary to severe intra-ocular disease, e.g., glaucoma, retinal separation, and infections.

As was mentioned in the chapter discussing the physiologic aging processes of the lens (page 1018) the nucleus gradually undergoes a process of hardening or sclerosis.* As part of these changes the relief of the adult nucleus becomes increasingly reflective and prominent. This serves further to help biomicroscopic distinction between the nucleus and the cortex. It should be remembered, however, that the zone or band forming the so-called "surface" of the adult nucleus is not a sharp one but in adults and older persons is rather an area of definite sagittal thickness, as evidenced by its rather cloudy and ill-defined demarcation when viewed in optic section. Evidently physiologic sclerosis is not strictly limited to the central parts of the lens but may also involve the deeper parts of the cortex. Except for the above mentioned phenomena, physiologic sclerosis *per se* does not result in opacification, at least of sufficient density to be recognizable biomicroscopically as a relucient haze. When a definite milkiess is found within the nucleus, we are already dealing with cataractous development.

Observation with the narrow beam shows that opacification of the nucleus occurs in a definite pattern and starts in a well-defined zone. The first indications of increase in relucency are seen within the two biscuit-shaped inner embryonal nuclei (Fig. 411 A, B). According to Koby, the anterior inner embryonal nucleus tends to become involved a little in advance of the posterior one, but Vogt states that he has never seen the anterior inner embryonal fetal nucleus opaque and the posterior one clear or vice versa. The dark central interval may remain recognizable for a long time (Fig. 411 B) but in the latter stages it may no longer be identified as such. An interesting and characteristic finding is that the anterior and poste-

* Vogt states that the widely held opinion that the as yet clear sclerosed nucleus lacks vital qualities and hence is to be considered as a kind of dead foreign body within the living lens is false. Nuclear sclerosis should be considered as a conserving process, which inhibits degeneration and in this sense tends to preserve vision for a longer time.

rior embryonal fetal sutures tend to become opaque, standing out as distinct, sharp, white figures, against the grayish-white (opaque) background of the affected inner embryonal fetal nuclei. The central

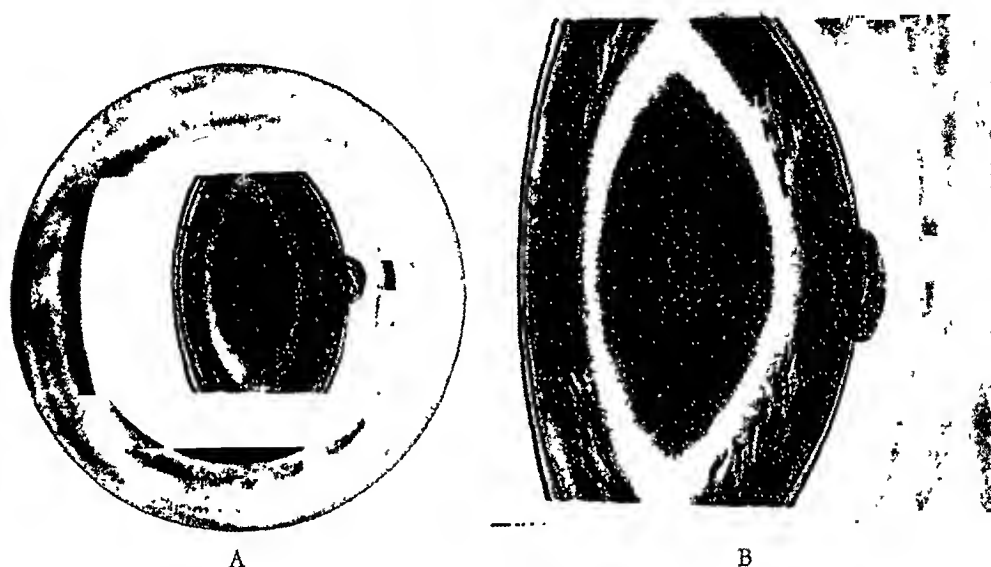


FIG. 411. Early nuclear cataract. A. The beam (optic section) passing through the lens is observed with the unaided eye. Note smaller posterior capsular opacity and faint cortical changes. B. Same as A, 15 \times magnification.

clouding progresses outwardly in both directions gradually involving both outer embryonal fetal nuclei. A characteristic finding at this stage, which was brought to our attention by Vogt, is the fact that the central opacity is always separated from the adult nuclear stripe by a dark (lucid) interval in all directions (Fig. 411 B). However, although this "lucid" interval may remain present for months or years, eventually as the reflection of the adult nucleus stripe increases, it becomes opaque so that in the end the central opacity merges with the highly reflecting adult nucleus stripe. As a result the nuclear cataract extends continuously from the anterior nuclear stripe to the posterior one. The opaque nucleus is then separated from the stripe of discontinuity by a more or less dark cortical band, depending of course on the relucency of the cortex which in turn is governed by cataractous changes within it.

In all cases, even with higher powers of magnification, the uniform opacification, causing the nuclear haze, cannot be resolved into any definite morphologic elements. Many writers, however,

have described a dustlike composition. This difference in behavior between the cortex and nucleus in this regard is striking. In the former gradual coagulation and liquefaction results in the formation

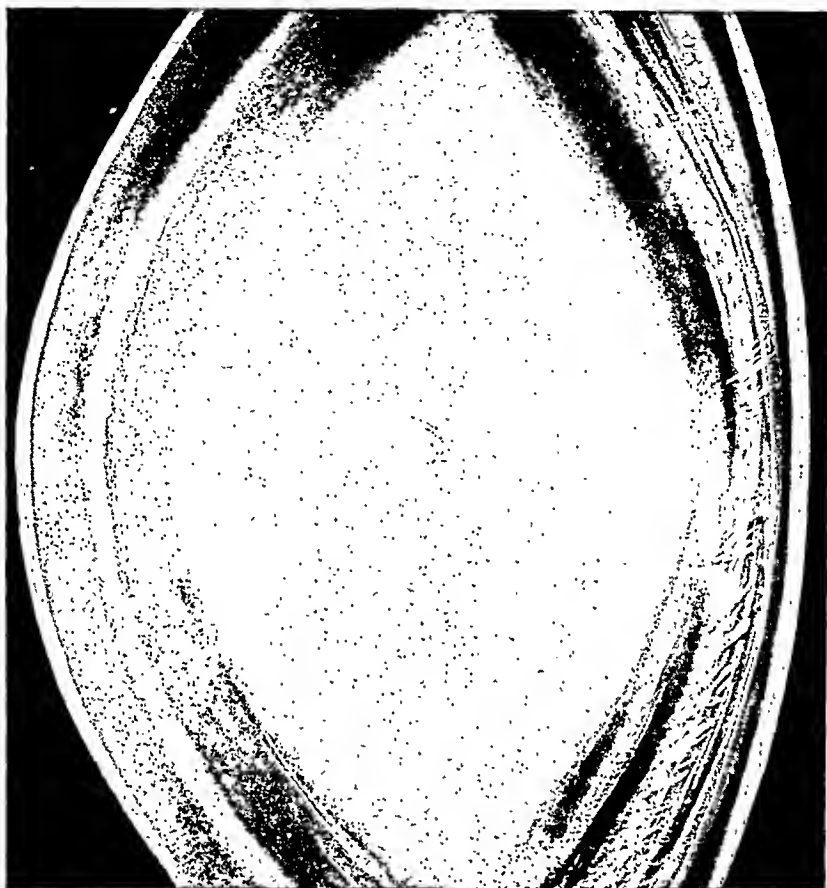


FIG. 412. Nuclear cataract further advanced; cortical changes are well developed.

of droplike elements with more intense opacification. As previously suggested this variation in the response between the cortex and the nucleus is probably due to differences in consistency (sclerosis of the nuclear fibers) rather than to any specific chemical change peculiar to either part. Because of the diffuse type of opacification common to nuclear cataract, it is possible with the narrow beam and sharp focusing to penetrate through the most opaque nucleus, provided the anterior cortex is not entirely opaque. It is now well known that nuclear cataract is closely associated with "lens of double focus" and is a forerunner of cataracta brunescens and nigra.

Nuclear Cataract with Double Focus. This is not so rare a phenomenon in senile cataract as thought; and has frequently been over-

looked. It now seems that, owing to a moderate opacification of the fetal nuclei (nuclear cataract) with retention of a dark surrounding interval, a difference in refractive index between the cortex and the nucleus results in a central myopia. There is a coexistent peripheral emmetropia, hypermetropia, or a smaller degree of myopia than that which occurs axially.

According to the records, this condition was first hinted at by L. Müller (1894),⁵⁴⁸ who published an observation of Salzmann concerning a lens with double focus. One year later, Demicheri³⁹⁸ described the condition in three patients and, because of its resemblance skiascopically to lenticonus, called it "false lenticonus." Following this report, several other authors made similar observations (Guttman,⁴⁶⁶ v. Szily,⁶²⁵ Halben,⁴⁶⁸ v. Hess⁴⁷²). Demicheri found that a severe disturbance of vision resulted from the fact that the axial parts of the lens showed a higher refraction (myopia) than did the periphery, the two being separated by an intermediary zone concentric to the lens surface. He concluded that the trouble lay in the difference between the indices of refraction of the cortex and the nucleus, as if a stronger refracting nucleus were located within a weaker refracting cortex. The fact that the inner nuclei were highly reflective (opaque) was not stressed by the earlier writers, perhaps because in many instances, the opacity was not visible ophthalmoscopically or only insignificantly so. But in 1903 von Szily with the method of oblique illumination at his command already spoke of the increase in inner reflection and suspected the presence of a cataract.

However, Vogt⁶⁵⁴ in 1923 stressed the fact that biomicroscopically the lens with double focus was dependent on nuclear cataract. It is found that the contours of the opaque part of the nucleus were more curved than those of the surface of the adult nucleus or capsule. In other words the great difference in index of refraction in the axial part is brought about by a pathologic cataractous process. It should be emphasized that the nuclear opacity is not total but only involves the fetal nucleus. The opaque fetal nuclei are separated

from the surfaces of the adult nucleus by a "lucid" interval. It will be seen that the condition of double focus can occur only with the average pupillary width provided the central axial opacity is sufficiently small (not over 4.5 mm.) to permit vision through it and at the same time through the unaffected peripheral parts around it. A form of double focus can also be demonstrated when the nuclear opacity is larger provided the pupil is dilated and a steneopaeic slit is employed. Occluding the central part of the slit it will be found that the distant vision is better (less myopic) than when direct central vision through the unoccluded slit is permitted. The fact that only in isolated cases does a lens with double focus occur, considering the great frequency of nuclear cataract, is substantiated clinically in that in most cases the nuclear opacification occupies a greater area. The further outward the opacification extends (i.e., toward the surface of the adult nucleus) the less curved will its surface be. Most cases of lenses with double foci occurred in older persons who were myopic. In these cases "progressive" myopia may occur within a few months.* These patients when looking in the distance seem to see better than their degree of myopia warrants owing to the fact that they employ peripheral vision. Among the other distressing symptoms are diplopia and polyopia. In lenses with double foci the nuclear opacification not only progresses with time, in which case the troublesome symptoms decrease, but also there is a strong tendency for them to develop color changes leading to cataracta brunescens and nigra. As the opacification develops the "lucid" area between it and the surface of the adult nucleus finally becomes relucet (Plate LXX, fig. 5). From the very onset the reflection from the surface of the adult nucleus is increasingly marked. Another feature, common to nuclear cataracts, is the appearance of the embryonal sutures, which stand out as white lines; also the fact that the dark interval, located between the inner fetal nuclei, may be identified as such in the early stages only. Later on it also becomes

* Vogt pointed out that eyes with axial myopia are predisposed to this condition. Likewise cataracta brunescens and nigra is relatively frequent in axial myopia as well as cataracta complicata secondary to intra-ocular disease. Lenses with double focus are rare in persons under 35 years of age, although he found one in a 10-year-old boy.

cloudy. Diabetes is another condition in which a rapid progressive myopia may suddenly develop. However, as Vogt has pointed out, diabetic myopia can occur without showing any definite or typical lens changes biomicroscopically. Also sudden myopia may occur during the course of sulfonamide chemotherapy.

CATARACTA BRUNESCENS AND NIGRA

One of the features characteristic of nuclear cataract is the frequently found color change in its deeper parts. As discussed on page 1012 under "physiologic" senile alterations, it is rare indeed not to find some yellow or yellowish brown coloration in the posterior subcapsular regions of the lens in persons over 60 years of age who still have normal vision. In studying such cases, I have found that frequently the yellow coloration begins at the posterior capsule and advances forward in the posterior cortex up to the surface of the posterior adult nucleus. Undoubtedly the "warm" coloration of the deeper parts of the lens acts as a filter and as such would affect vision in a way similar to tinted spectacles. Any greater reduction (in the absence of other changes) would have to be attributed to the associated nuclear opacity. At any rate, in many cases of nuclear cataract, further extension of this change results in the appearance of a tan, orange, or red to brownish-red color (Plate LXX, figs. 1, 2, 3, 4). The terms "cataracta brunescens," "rubra," and "nigra" are employed to express the varying degrees of color intensity seen. In the extreme state, ophthalmoscopically, we find a black, nontransilluminable pupil (the so-called "cataracta nigra").* It should be pointed out that this intense coloration, although associated with it, is probably not entirely dependent on nuclear sclerosis and cataract alone. Even in the presence of complete nuclear opacification, the reflex from the deeper parts may only show a weak yellow color, not unlike that seen physiologically in age. The color change which is most marked in the posterior cortex gradually extends forward until

* With the moderately narrow beam, however, in this instance it will be seen that the color is an intense dark red, increasingly so as one approaches the deeper parts. With diffuse illumination it is evident to a lesser degree and consequently the posterior half of the nucleus will appear darker or blackish. Red-free light does not penetrate at all and the posterior parts will be jet black.

PLATE LXX

FIG. 1. Nuclear cataract (brunescence). Direct focal illumination. Low power.
(After Messmann.)

FIG. 2. Nuclear cataract. Direct focal illumination. Optic section.

FIG. 3. Nuclear cataract (Rubra). Direct focal illumination.

FIG. 4. Nuclear cataract. Direct focal illumination.

FIG. 5. Nuclear cataract. Optic section. Direct focal illumination.

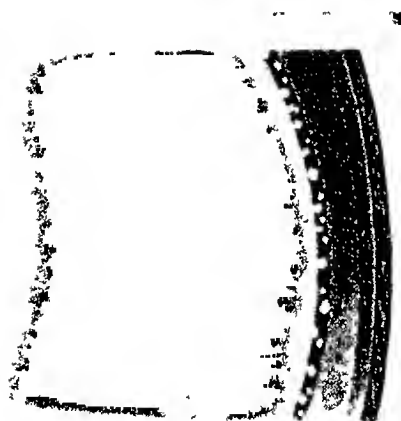
FIG. 6. Nuclear cataract with central area of unusual vacuolar degeneration.
(Courtesy of Dr. D. Gordon.)



2



1



3



4



6



5

it fades within the nucleus. The most anterior parts of the adult nucleus and the deeper and middle anterior cortex frequently have a decided greenish-gray tinge, in contrast to the bluish-gray one seen normally. Thus, within a single lens, we may find a gradual "dilution of color," varying posteriorly from a dark red or brown to a light yellow and greenish tinge. As the process develops into a definite cataracta brunescens, rubra, or nigra, the brownish or deep red color extends itself almost to the anterior adult nuclear surface where it changes into orange and then to greenish-gray. Only the anterior sub-capsular region (in the absence of cortical changes) may still retain the original bluish-gray opalescence characteristic of the normal lens. Vogt postulates that color variations are simply an expression of dilution of one type of dye stuff. He compares it to the color changes seen with gradual dilution of certain colored chemical substances, e.g., lysol. In concentrated form a thin layer of lysol solution appears red to red-brown while a thicker layer of the same concentration is black. Highly diluted, it is pale yellow. However, Bellows refers to the work of Walls and Judd (1933)⁶⁷⁰ and Walls (1931, 1940)^{668, 669} on the coloration of lenses in lower animals. They found that in certain animals whose lenses were yellow the intensity of this color was related to the degree of exposure of these animals to bright light. They were able to extract a yellow pigment which they called "lenti-flavin" and which apparently differs from the pigment in cataracta brunescens and nigra.

Although the fainter yellow coloration occurs in practically all persons of great age especially those with senile myopia, well-developed cataracta brunescens and nigra, according to all available statistics, is rare. Rollet and Bussy,⁵⁸¹ gathering data from several authors, found it 48 times in 15,763 cases of cataract extractions. Also coloration of both lenses in the same individual can differ markedly normally or in cases where severe disease of the inner eye exists (cataracta complicata).

For a long time there has been considerable dispute concerning the mechanism producing these color changes, especially whether it is derived from extra- or intralenticular sources and whether it is

caused by purely physico-optical phenomena or by the actual production of pigment.* Suffice to say here that modern opinion holds that the coloring represents pigment formed endogenously and that it probably has a "melanin-like structure" (Bellows). It is interesting to note that in one place in his atlas, Vogt follows other writers (Hess, Busacca and others) in favoring the physical theory for the explanation of the color and in another he speaks of the presence of a dyestuff (i.e., the color pigment may be formed by a transformation of the lens proteins). The current theory is that melanin or "melanoid-like substances" are derived from a precursor, tyrosine. Since free tyrosine is only found in cataractous lenses, it follows that decomposition of lens protein is a prerequisite. In a similar way conjunctival and corneal pigmentation (Stahli's line) may be a result of the liberation of premelanotic substances in the pathologically changed protein content of these tissues (see Vol. I).

* For a more complete discussion of this problem the reader is referred to other sources, e.g., Hess, 1911;⁴⁷² Rollet and Bussy, 1921;⁵⁸¹ Vogt, 1931;⁶⁶² Gifford and Punttenney, 1933;⁴⁴⁸ and Bellows, 1944.³⁶⁴

COMPLICATED CATARACT

IN ITS broadest sense, complicated cataract (*Cataracta Complicata*) refers to lens changes incident to the action of certain toxins. These toxins may originate within the eye itself (endogenous) or may be derived from extra-ocular (exogenous) sources. The exact mechanism by which they act is not known but it is conjectured that they diffuse into the lens from the surrounding fluids and cause alteration in its metabolism. Hence they differ from developmental cataracts (congenital, presenile and senile) which in a sense are predestined either by hereditary influences or are expressions of abiotrophy or senility (the appearance of which may also be genetically determined even in isolated tissues or organs).

Although the characteristic picture of this condition is predominantly found in the region of the posterior capsule, anterior subcapsular changes occur as an expression of complicated cataract more frequently than has hitherto been considered. The appearance of *cataracta complicata* may vary depending on the location of the causal factor. For instance, in certain forms of exogenous complicated cataract (e.g., tetany, myotonia, diabetes, radiational cataracts), we find subcapsular layers of opacities (anteriorly and posteriorly) which may spread out over the entire lens surface. The anterior changes consist of capsular thickenings and opacities while subcapsularly thin layers of vacuoles are seen. These may be flat bands of rosette-like opacities, isolated spots and layered roundish or punctate changes. Although one might like to think that anterior complicated cataracts uniformly are associated with conditions in the anterior segment of the eye, and that posterior complicated cataract strictly follows disease within the posterior segment, actually this

is not so. Cases having posterior complicated cataract sooner or later show anterior subcapsular changes. Especially common is the development of nuclear cataract. In this connection it should be pointed out that in high grade myopia and retinal separation a nuclear cataract indistinguishable from the ordinary senile variety, may be the only visible cataractous change present.

One of the great advances afforded by biomicroscopy of the lens is the ability to differentiate between ordinary senile cataracts and cataracta complicata. This is a consequence not only of their morphologic differences but also because we are better able to see changes (Inflammation) in the neighboring tissues, which may be the basis of cataracta complicata.

To the original conception of Becker (1876)^{359, 360} — i.e., that complicated cataract follows such diseases as retinal detachment, intra-ocular tumors, cysticercus, absolute glaucoma, cyclitis, iridocyclitis, and the unknown processes, which result in buphthalmos — Fuchs (1910)⁴³³ added high-grade myopia and violent suppurative diseases of the cornea (ulcus serpens). In addition, now, are included the exogenous endocrine cataracts (diabetes and tetany, myotonia, mongolian idiocy, and the dermatoses, etc.), radiational cataracts, and cataracts following intoxications (drugs, such as naphthalene, dinitrophenol, paradichlorobenzene, thallium, ergot, etc.) and also certain retained metallic foreign bodies (iron and copper) producing siderosis or chalcosis lentis. However, since many of the last-mentioned varieties have special characteristics, they will be treated under special sections. Although from the standpoint of etiology, all these forms fall under the heading of complicated cataract and seem to have a predisposition for the subcapsular regions, anteriorly as well as posteriorly, those which are secondary to intra-ocular disease (endogenous) start chiefly axially, in the posterior cortex and usually show color display and a porous consistency.

Various suggestions have been offered to explain the reason why complicated cataract, especially following intra-ocular disease, occurs predominantly in the posterior subcapsular regions. Among these is the fact that the thinnest part of the whole lens capsule is

the axial portion posteriorly. In addition the absence of protective epithelium posteriorly should be kept in mind. Both these factors might conceivably offer a weaker barrier to the entrance of toxins in this part of the lens. Also to be considered is the point that the posterior portion of the lens lies in closer proximity to the sites from which noxa emanate. However, in my opinion the difference in the chemistry and physiology between the aqueous and the vitreous in this regard should be stressed. On the one hand there is a watery solution, capable of being more or less continuously renewed. On the other hand, there is the torpid vitreous gel. This significance is exemplified in the thoroughness and speed with which the aqueous cleanses itself of toxins and exudates as compared to the vitreous. The suggestion of Vogt that the sutures are places of less resistance and consequently are sites of predilection as proved by the frequent rosette-like formations seems to me to be well taken since most complicated cataracts assume a radiating design. The close relationship between retinal pathology and the integrity of the lens has been noted by numerous writers.* The frequency of posterior complicated cataract in retinitis pigmentosa, high-grade myopia with chorioretinitic changes, as well as retinal separation bespeaks this relationship.

BIOMICROSCOPIC APPEARANCE OF CATARACTA COMPLICATA

Anterior Form. This occurs not uncommonly as a consequence of iridocyclitis. The lens opacity is usually to be found subcapsularly, axially or paracentrally, in which case it tends to lie in the vicinity of iritic adhesions or in the region of maximum iritic involvement (Plate LXXI). The cataract itself may be localized to one area or may be spread out in the form of thin flat bands to assume a rosette-like figure. In the early stages (*cataracta complicata incipiens*) the opacity, which can be missed by ophthalmoscopic examination, appears in direct focal light as a faintly grayish haze.

By diffuse or retro-illumination and higher powers of magnifica-

* Spemann,⁶¹⁵ Lewis,⁵²⁰ Stockard⁶²⁰ and a host of other workers in the field of experimental embryology have shown in lower animals, the dependence of the development of the lens on that of the optic cup. It is conceivable that in mammals, if there was a weakness of genetic origin in the ectodermal epithelium of the optic cup, it might also affect the lens.

PLATE LXXI

FIG. 1. Anterior complicated cataract following contusion. Capsular folds. Diffuse illumination.

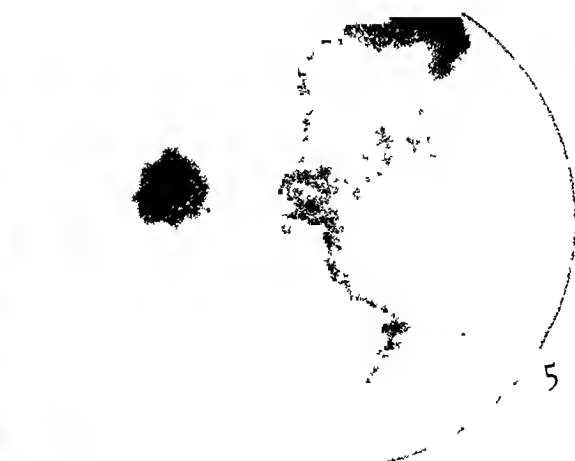
FIG. 2. Same as case shown in Figure 1. Direct focal illumination. Optic section.

FIG. 3. Posterior complicated cataract (endogenous). Following iridocyclitis. Diffuse illumination.

FIG. 4. Same as shown in Figure 3. Direct focal illumination.

FIG. 5. Complicated cataract (nonprogressive) several years following a perforating injury. Located at the lens equator.

FIG. 6. Unusual form (lamella) of anterior and posterior complicated cataract located at the level of the adult nuclei.



tion the opacity is seen to be composed of delicate drops or vacuoles (Fig. 413). In other instances it is composed of whitish or grayish points (Fig. 416 B). At times the opacity may be circular (ring-

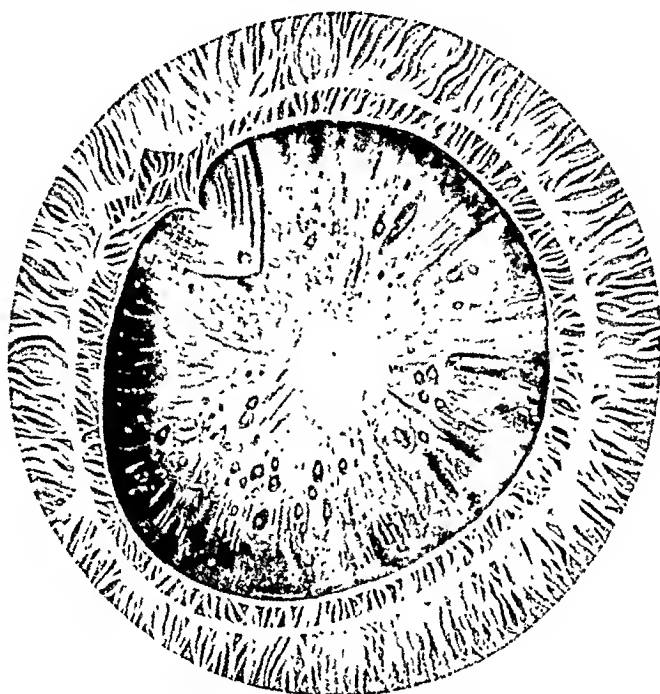


FIG. 413. Frontal view by diffuse illumination. Anterior complicated cataract, vacuoles and opacities. Posterior involvement not seen.

form) with or without delicate radiating branches. Apparently depending on the length of time of action of the influences, these opacities may be stationary for long periods of time or may progress.* With progression they become more prominent and tend to expand in the direction of the sutures and at the same time to extend themselves deeper within the cortex. Eventually the whole cortex may be involved so that it becomes entirely opaque, made up of a vascular and milky substance. With hypermaturity, similar to senile cataract, capsular folds and calcareous degeneration occur and finally the entire lens may shrink into a small irregular yellow mass.

Posterior Form. As mentioned before, the characteristic posterior cataracta complicata may follow iridocyclitis as well as degenerations

* In one case of healed iridocyclitis Vogt thought that after 9 years three ring-shaped subcapsular opacities with fine radiating stripes were unchanged. In another healed case, 11 years later, the opacity had not progressed but was more distinct and was located somewhat deeper in the cortex. Apparently like other localized cortical opacities, it was dislocated backward by the ingrowth of new fibers.

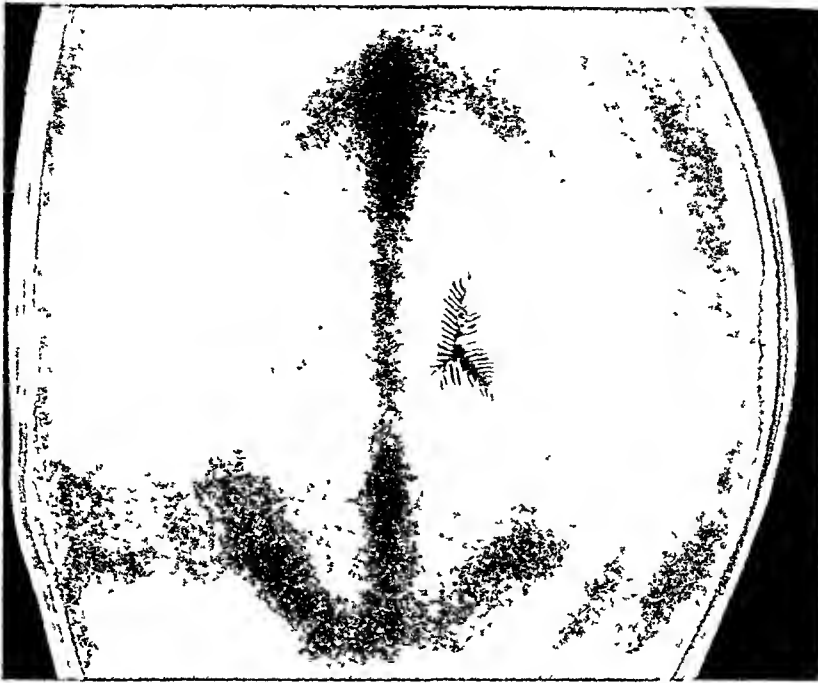
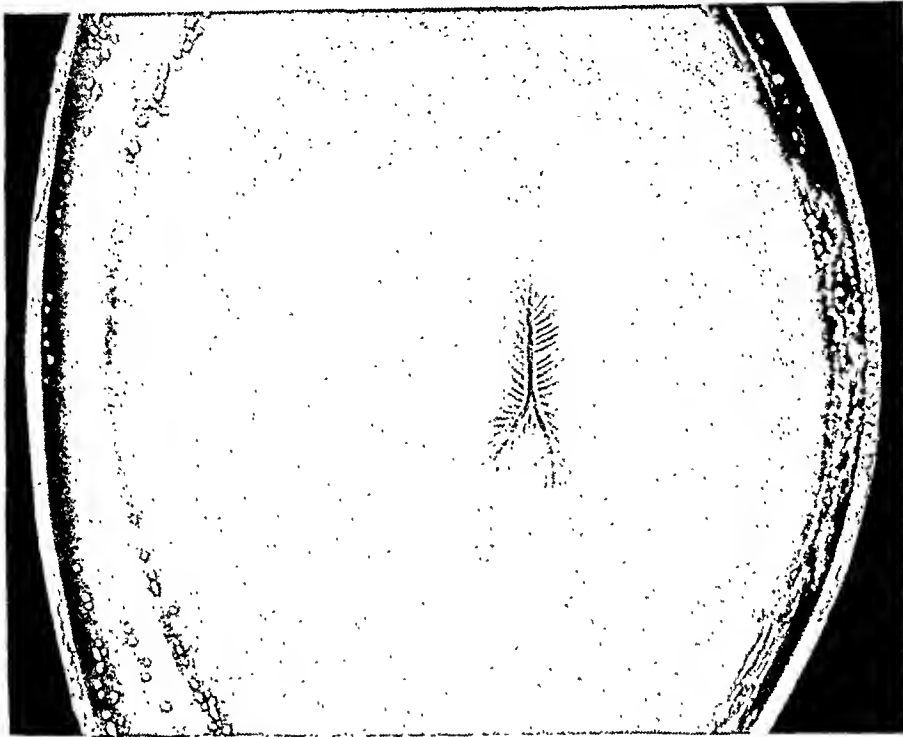


FIG. 414. Anterior and posterior complicated cataract seen in optic section.

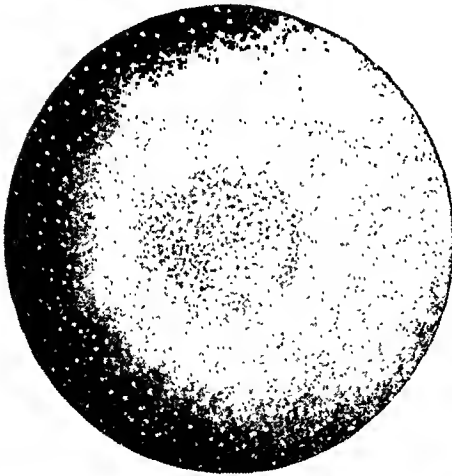


FIG 415. Early cataracta complicata (uveitis). In the region of the bright specular reflex (above) a definite color display (iridescence) was noted. A few pigment deposits were seen on the posterior lens capsule.

(retinitis pigmentosa and retinal detachment) and inflammations of the posterior segment. Any and all types of conditions may occur, e.g., tumors seem to predispose to this form of cataract but not in



A



B



C

FIG. 416. A. Early stage of posterior complicated cataract (uveitis). Optic section. B. Note rounded form of opacities (frontal view). C. Cataracta complicata (posterior) associated with retinitis pigmentosa.

every case since nuclear cataract and posterior saucer opacities are also found with these conditions.

Typically complicated cataract begins as a localized hazy opacity

adjacent to the posterior capsule and usually in the polar region (Fig. 415; Plate LXXI). It may, however, start paraxially or even peripherally. It should be pointed out that although in most cases the

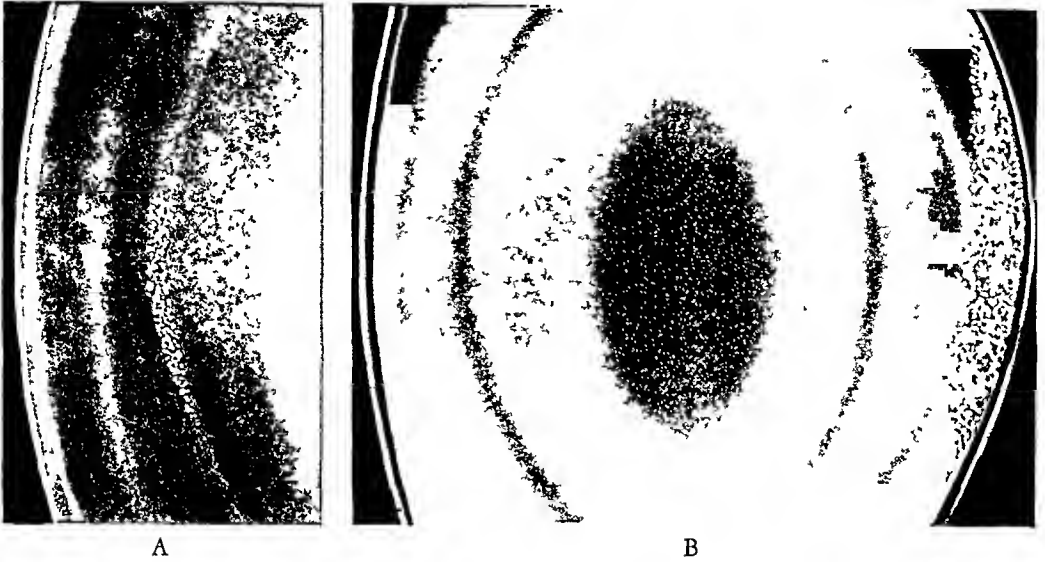


FIG. 417. A. Complicated cataract associated with retinitis pigmentosa located in the anterior part of the posterior cortex. B. Posterior complicated cataract associated with retinitis pigmentosa as observed in optic section.

cloudy opacification at its inception seems to be connected with the posterior capsule or just in front of it, rarely it may lie separated from the capsule in the posterior cortex (Fig. 417 A; Plate LXXI, fig. 6).

With higher powers it will be seen that the opacity is composed of coarse granular particles within a hazy media. These particles (probably brought about by the formation of vacuoles) have been described as porous in character (like pumice or tuff-stone) and by others as resembling bread crumbs or chopped meat. Frequently, larger ringlike opacities have a dark, more or less optically "empty" content (Fig. 416). This can be demonstrated by optic section. These structures represent fluid spaces surrounded by opaque borders and are also often seen in anterior complicated cataract. Unlike water slits or laminary separation they do not follow the direction of any of the lens structures (sutures or fibers). As a matter of fact this apparent "lawlessness" is also characteristic of expansion of the opacification in all types of cataracta complicata. In cases of retinitis pigmentosa, a typical form of very slowly progressive complicated

cataract is found (Fig. 417 A, B; Plate LXXVI, fig. 6). It frequently is irregularly roundish or has a starlike shape with three or four radiating processes and is composed of crumblike particles or of a small

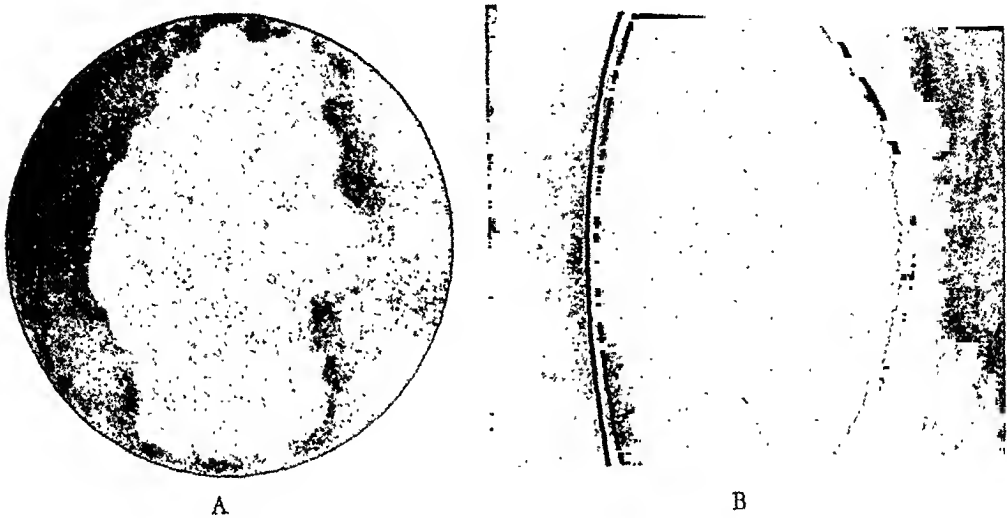


FIG. 418. Complicated cataract. A. Diffuse view. B. Direct focal illumination.

ringlike vacuolar net, the centers of which are less opaque (Figs. 416 B, C). Complicated cataract of a similar morphology is also found in the higher grades of myopia (myopia degenerativa), especially in the presence of vitreous and chorioretinal changes (Fig. 418 A, B). This type of change in degenerative myopia should be differentiated from posterior saucer-like opacities, which are often found in myopes of advanced age and which probably represent a senile change independent of the myopia. The same possibly holds true for the frequent finding of nuclear cataract in older myopes.

An early finding in cataracta complicata is the presence of a brilliant color display (red, yellow, green, and blue) in the affected area, especially as the zones of specular reflection are approached (Plate LXXII, fig. 6). Our attention to this polychromatic display was called first by Vogt who placed much importance on it as a diagnostic feature of cataracta complicata.* This color display or iridescence occurs not only in axially located opacities but also in peripheral ones. He believed that it has its origin in a very thin layer of subcapsular fluid, the thickness of which could not be over 0.2μ .

* I have frequently seen a similar color display in cases of posterior saucer cataract.

After progression and thickening of the opacity, evidence of iridescence may be absent. A marked color display was noted by Vogt in cases of recent retinal separation with incipient but rapidly progressive cataracta complicata. With the ophthalmoscope many flat vacuoles were seen. Biomicroscopically they were localized just in front of the posterior capsule. The opacification at first was of little density but later the edges of these fluid spaces developed whitish porous opacities, and the typical rings appeared. The vitreous, as is common in these cases, showed a reddish-brown cellular infiltration and changes of structure. With development, and especially in older persons as is the case in posterior saucer cataract and nuclear cataract, the color of the opacities in this region may become increasingly yellow.

The porous structure of posterior complicated cataract differentiates it from the changes usually found in cortical cataract — water slits, spokes, etc. (Fig. 419). The opacity tends to advance in two directions, spreading rosette-like to the periphery in the direction of the sutures so that the whole posterior subcapsular area of the lens becomes involved (like saucer cataract) and then spreading sagittally in the direction of the posterior adult nuclear stripe. The latter extension results in a considerable thickening of the opacity, especially in the polar regions. This type of irregular anterior extension into the posterior cortex differentiates complicated cataract from senile posterior saucer cataract whose anterior surface is sharply demarcated, since in this form of cataract progression forward usually does not occur. However, in the early stages, particularly in the absence of degenerative and inflammatory signs in the neighboring parts (which is rare) and based upon morphologic aspects alone, it may be difficult at times to differentiate between them. Posterior traumatic rosette opacities (page 1250) may occasionally present a problem in differential diagnosis. These, like posterior-saucer cataract (which ordinarily does not assume the rosette form) are sharply demarcated. But traumatic rosette differs from both saucer-cupiform cataract and complicated cataract in that it tends to be non-progressive.

With development, the outspoken characteristic of cataracta com-

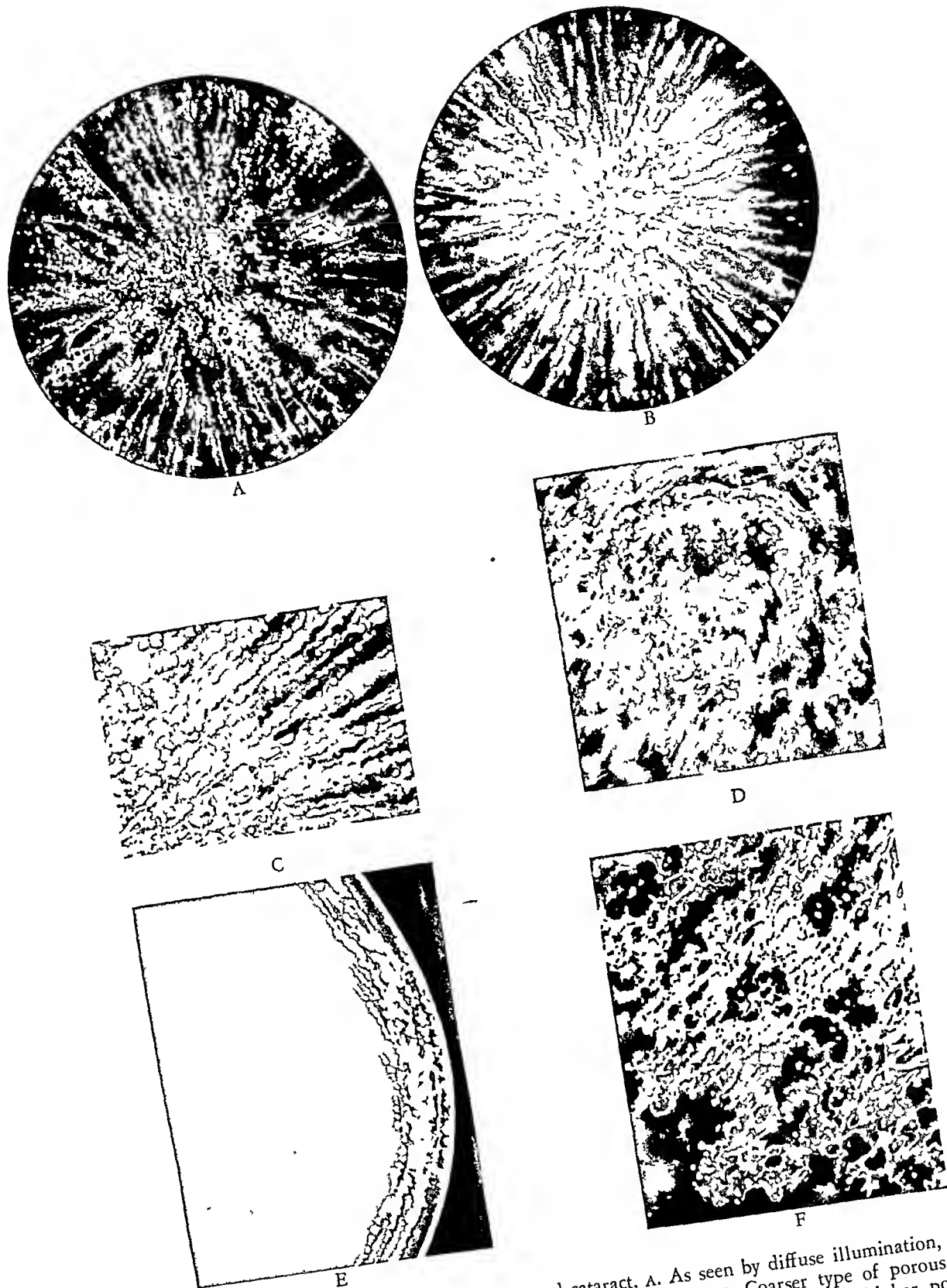


FIG. 419. "Porous" opacities of complicated cataract. A. As seen by diffuse illumination, low power. B. Another case, showing details under higher power. C. Coarser type of porous-like opacities as seen by diffuse illumination, low power. D. Same case as in C, higher power. E. Optic section through porous-like opacities. Note the extension of the opacities into posterior cortex. F. Details of the opacities shown in E under diffuse illumination, high power.

plicata becomes so obvious that its identity becomes unquestioned even to the beginner. The axial parts of the posterior cortex become filled with the large whitish crumblike irregular condensations which are especially thick in the polar regions, and from this radiating bands of the same porous nature extend toward the periphery where they become progressively thinner. These white crumblike structures within the opacity are suggestive of calcareous degeneration. The whole figure lies within a hazy medium. At times the opacity may become separated from the posterior capsule and may approach the posterior adult nuclear stripe. Within the central parts Vogt found ring forms and occasionally in the region of the posterior pole two or more ill-defined layers of opacity, one in front of the other, in which the anterior layer was the denser. Anterior subcapsular changes may appear early or late. Even in the early stages of cataracta complicata it is not unusual to find a few round vacuoles of varying sizes beneath the anterior capsule. However, these are often seen in senile cataracts as well. Later, fine flat subcapsular opacities appear; by retro-illumination and higher power these will be found to be composed of the finest vacuoles, the onset of gradually progressive anterior complicated cataract. Likewise, the appearance of nuclear cataract is common in the later stages.

Eventually, if the process advances sufficiently, in which case it is usually associated with progressive deterioration of the eye as a whole, a state of hypermaturity or shrinkage results (not unlike that of senile cataract). This is particularly so in long-standing chronic iridocyclitis and absolute glaucoma in which the pupil becomes secluded and the iris very atrophic. The capsule becomes whitish and thickened by deposits and filmlike exudative membranes, and tends to develop the characteristic double reflecting folds. These folds may extend across the lens surface in parallel or crossed lines and at times may be outlined by pigment deposits. With extreme shrinkage of the lens, parts of a black pupil may become visible and occasional zonular fibers may be seen stretched across it. Within the hypermature cataracts, calcareous changes and deposits of cholesterol crystals may be found.

CATARACTS ASSOCIATED WITH ENDOCRINE DISORDERS

Clinically there is considerable available evidence that endocrine dysfunction may lead to the development of cataract. Considering the present-day lack of information concerning the function and interrelationship of the ductless glands, it is not surprising to find confusion in the literature concerning the so-called "endocrine cataracts." Bellows states: "Only in diabetes and parathyroid tetany is there a very solid foundation of experimental data to support the clinical observations. The cataracts observed in albinism, mongolian idiocy, gonadal insufficiency, myotonic dystrophy, neurodermatitis and scleroderma, although presumably endocrinal in character, have little experimental confirmation." Even from the clinical standpoint, a great deal of statistical data is still needed. These will have to be coordinated with more exact morphologic descriptions of the changes, now possible with the biomicroscope.

At any rate, in these conditions (not dissimilar to complicated cataract in general) the opacities are found predominantly in the subcapsular middle portion of the anterior and posterior cortex, in most cases leaving the zones of disjunction unaffected. The appearance of the component parts of the opacities varies in individual cases but characteristically they are composed of whitish, discrete, fine, powdery, punctate dots, larger angulated spots, irregular flakes and occasionally iridescent opacities and crystals. In any one case, one type may predominate or several different forms of these constituents may be found to coexist in varying proportions. In addition thin flattened subcapsular vacuolar opacities spreading over a considerable area, not unlike those described in anterior complicated cataract (page 1161), may occur. It should be emphasized that, despite their characteristic location and appearance, all these types of opacities are not pathognomonic for any specific endocrine disorder. Similar ones may occur in other conditions, e.g., those resulting from the effects of radiant energy, toxins (drugs), and trauma. A point to be stressed is that apparently in most complicated cataracts the youngest

PLATE LXXII

FIG. 1. Diabetic cataract showing layers of anterior and posterior punctate opacities. 12-year-old girl. Direct focal illumination. Low power.

FIG. 2. Same case as shown in Figure 1. Direct focal illumination. Optic section. High power.

FIG. 3. Diabetic cataract (rosette form). Woman aged 28.

FIG. 4. More advanced opacities in the diabetic cataract. Note dense central subcapsular opacity and cortical water clefts. Diffuse illumination.

FIG. 5. Same case as shown in Figure 4. Direct focal illumination. Optic section. Passing through the edges of the dense central opacity showing its subcapsular location. Cortical water clefts are not shown.

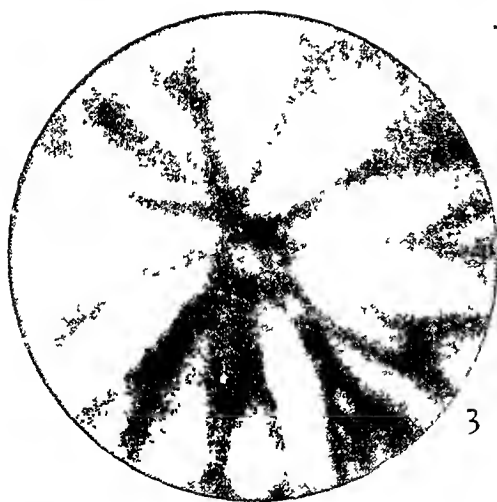
FIG. 6. Posterior subcapsular opacities (diabetic cataract). Note color display in area of specular reflex and beginning nuclear haze.



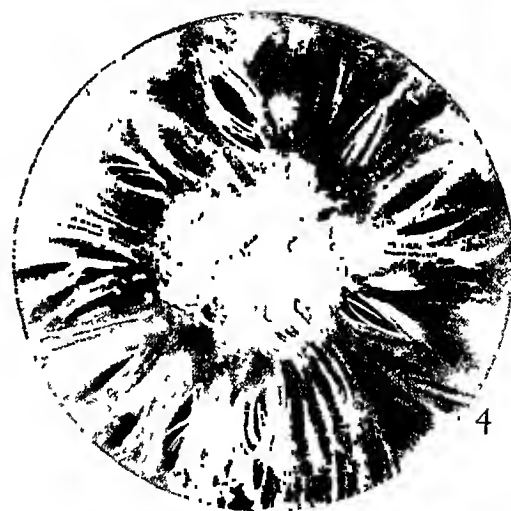
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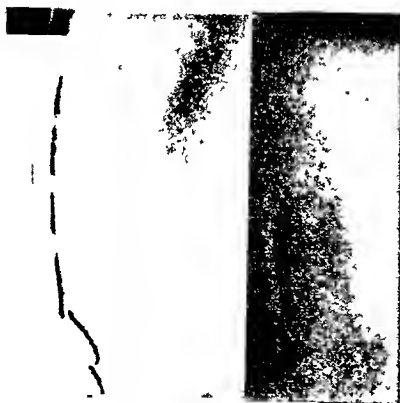
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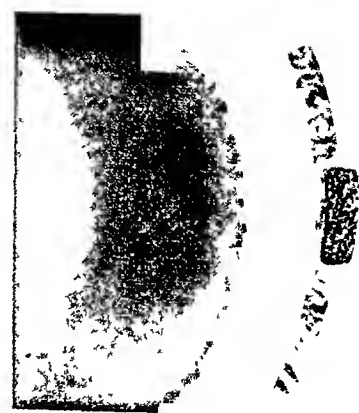
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5



6

subcapsular fibers of the lens seem to be vulnerable. However, one cannot deny the frequent occurrence of nuclear cataract (indistinguishable by itself from the common senile variety) in cases in which cataracta complicata would ordinarily be expected, but this form — with the possible exception of the diabetic cataract in older persons — is not typical for endocrine cataract.

As Duke-Elder has indicated, the striking clinical characteristics of endocrine cataracts in their pure form, although they frequently are complicated by other types, are the early onset, the bilateral incidence, and the zonular (layered) distribution.

DIABETIC CATARACT

At the present time the only type of cataract that justifies the appellation "diabetic cataract" is the rapidly progressive one that is found in younger persons (under 40 years of age) and that at the outset is characterized by the presence of a layer of more or less dense small punctate or flaky opacities involving the anterior and posterior cortex beneath the capsule (Plate LXXII, fig. 2). There is not sufficient experimental or clinical evidence at hand to evaluate how much of a role diabetes plays in the appearance of cataract (which clinically differs in no way from the ordinary changes of senile cataract) in the older age groups. As has been pointed out, a large percentage of apparently normal persons over 60 years of age show indications of lens changes of one kind or another. Therefore it is not surprising that in the past large numbers of ordinary senile cataracts were classified as "diabetic" just because they were found in older persons having the disease. Also in these cases there seems to be no relationship between the duration or severity of diabetes and the onset or progress of the cataractous changes. However, as suggested by Vogt, it could be supposed that, in instances in which a person is "genetically predisposed" to senile cataract, it could be provoked prematurely by diabetes. This author cites other conditions, e.g., senile corneal lines, arcus senilis, and coronary cataracts, as examples of conditions that may be prematurely provoked by outside influences (inflammations or toxins). Also to be considered is the idea that,

owing to the physiologic alterations (sclerosis, etc.), the "aged" lens may be incapable of reactions morphologically similar to those occurring in the young. So that the same exogenous influences acting in the young might produce dissimilar pictures in the aged. Hence, in the aged there is perhaps a tendency for even cataracts caused by exogenous toxins or even trauma to approach in appearance those of the so-called "ordinary" senile cataracts. Many theories have been advanced concerning the mechanism with which cataract could develop as a consequence of diabetes. Bellows summarizing the experimental evidence states: "Most writers attribute the opacity to osmotic or toxic actions of the excessive glucose and its metabolic products." On the other hand, recently it has been indicated by several workers that many persons (not frankly diabetic?) with ordinary senile cataracts have an inadequate mechanism for the utilization of sugar, and that "the incidence of hyperglycemia and decreased tolerance for carbohydrates was much greater than could be anticipated from senile changes alone."

Only those cases that occur in younger individuals (under 40 years of age) and that in many ways are morphologically similar to those found in other endocrine disturbances can be considered as true diabetic cataracts. Evidently the younger the diabetic person is in whom cataract appears, the more certain it is that we are dealing with diabetic cataract. Two chief features are to be noted in true diabetic cataracts: (1) their rapid development (usually bilateral) and (2) the subcapsular location of the opacities. Cases have been reported in which the opacification appeared within hours or days, and in one instance the lens became entirely opaque within 48 hours. This rapidity of development is similar to what occurs in certain toxic cataracts (page 1199). However, exceptions may be found. In the case of a 14-year-old diabetic girl (referred by Dr. Nemerson) on whom I performed a linear extraction, 4 years elapsed between the onset of the opacification and the time when operation became necessary. Her vision was reduced to 20/200 bilaterally. Forty units of protamine zinc insulin daily kept her sugar-free but apparently did not protect her from the slow development of the cataract. The

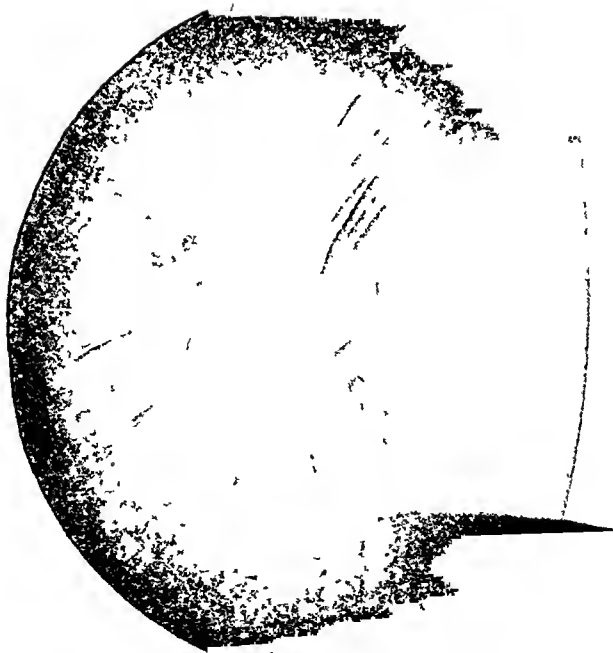
opacities in the beginning resembled the typical anterior subcapsular "snowstorm" cataract described by O'Brien. These consisted of dots and flakes varying in size in one eye, the smaller ones seeming to collect just behind the capsule but in this particular case they did not invade the space between it and the anterior stripe of disjunction* (Plate LXXII, figs. 1, 2). About a year later the opacities extended themselves in a gradual way into the middle and deeper parts of the cortex. Still later, posterior cortical opacities and, finally, a generalized nuclear haze ensued. The earliest changes seen by the writer occurred in a 28-year-old woman (Plate LXXII, fig. 3). They consisted of a single layer of glistening rosette opacities at the level of the anterior and posterior lines of disjunction. They were most marked in the peripheral regions and gradually faded out axialward. The rosettes were composed of shining dots which could only be seen by means of the optic section. They were invisible in strong diffuse illumination. Several months later, no change in their appearance was noted.

Vogt described the case of a 33-year-old woman in great detail. Ophthalmoscopically an opacity extending over the whole lens surface was seen consisting of small whorls and stripes. Also by decentering the light as it transilluminated the pupil, many small spokes became visible. Biomicroscopically in optic section both lenses appeared thinner than normal in sagittal direction, but in the left lens the cortex seemed to be thicker than normal. The zones of discontinuity were poorly outlined. The mass of opacities was located under the anterior and posterior capsule centrally and peripherally as far as it could be inspected. The opacities anteriorly were gray, cloudy and transparent while posteriorly they were flat, white, and compact, differing in this way from the porous opacities of posterior cataracta complicata. In the left eye the posterior opacity in the region of the pole measured about 1 mm. vertically and 2 mm. horizontally. While in the right (Fig. 421) the denser posterior ones were from 0.2 to 0.3 mm. in diameter. Elsewhere the subcapsular

* This differed from Goulden's observation in that he found that this space is usually involved.⁴⁵⁶ When the space between the capsule and the anterior stripe of disjunction is invaded, granular opacities may project into it and obliterate the stripe. When this occurs fluid (vacuolar) may occupy the area between the capsule and these opacities.



A

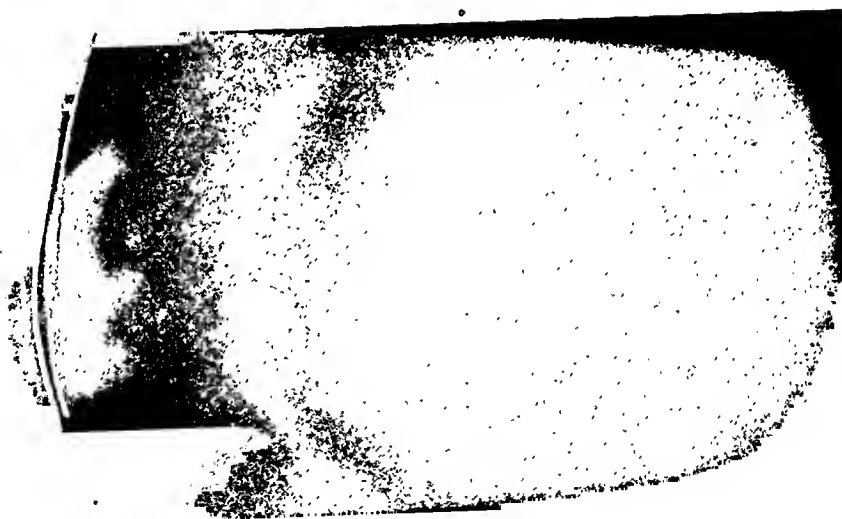


B

FIG. 420. Diabetic cataract with starlike figures. A. Opacity seen by diffuse illumination.
B. Same as A, as seen by retro-illumination in light reflected from the posterior capsule.



C



D

FIG. 420. C. Star-like figure (diabetic) by diffuse illumination. D. Same case as C, viewed in optic section. Note the location of the star-shaped figure at the level of the anterior line of disjunction. Also observe the beginning of a nuclear cataract.

opacities were mostly dot- or dustlike and invaded the deeper parts of the cortex. Occasionally colored glittering crystal line structures were seen in the neighborhood of the anterior mirror region. Vogt

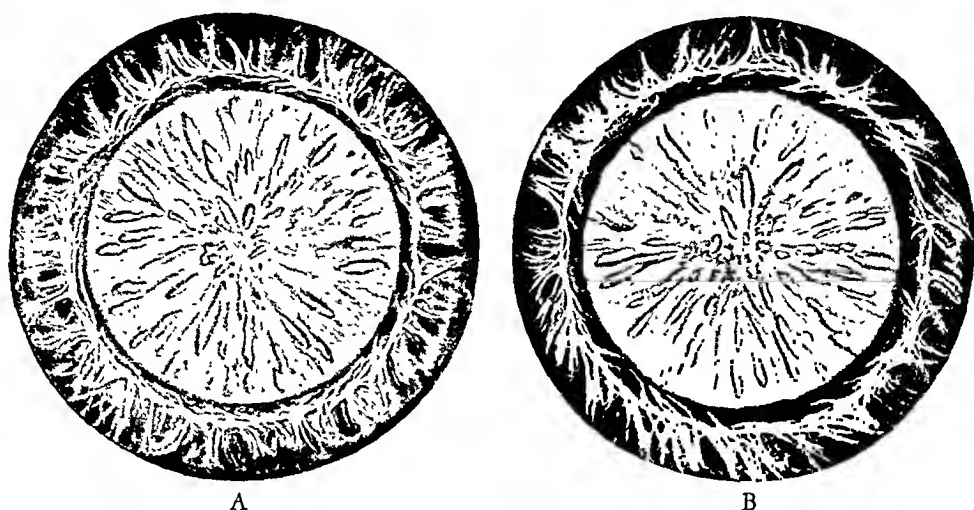


FIG. 421. Diabetic cataract. A. Right eye. B. Left eye. (After Vogt.)

also noted the appearance of opacities outlining the superficial fiber design. These speak for the involvement of the superficial cortical fibers. In places they were interrupted by lanceolate gaps and in the deeper cortex traces of water slits were seen (Fig. 421.)

As the formation of the fiber design develops, cortical vacuolization and opacification increases so that in the end the subcapsular punctate opacities are replaced by radiating opaque granular masses separated by irregularly dark lancet-like spaces. This picture, which is especially characteristic in the frontal view by diffuse illumination, can also be seen against the reddish glow of the fundal reflex ophthalmoscopically. By the latter method illuminated gaps (water-slits) between irregular darker outlines (radiating opacities) are visible. It should be pointed out that subcapsular fiber design can also occur in complicated cataracts from other causes (e.g., tetany, myotonia, trauma, and toxins). It merely illustrates the fact that in cataracta complicata the youngest fibers are the most vulnerable. Localized, subcapsular star-formed opacities may occur especially in the polar region (Fig. 420; Plate LXXII, fig. 3). In a recent publication, Rosen⁵⁸⁴ considered that the presence of the so-called

"diabetic needle" or "Roman numeral sign." * According to Rosen, this sign is not constant but, when present, is specific for diabetes. He states that, "Characteristically when the pupil is dilated and when the red reflex is studied with the indirect ophthalmoscope or retinoscope, a spoke formation is seen in the periphery of the lens, which is a linear black streak no thicker centrally than it is peripherally. This is its most important feature — the spoke is never thicker at the periphery than at its central termination and even in cases where there is adult spoke formation in the lens characteristic of early change in senile cataract, still there may be superimposed upon this cataract formation, the characteristic diabetic needle."

In his description and illustrations Rosen does not give any bio-microscopic descriptions of these "needles."

In the main, Vogt emphasized that the characteristic picture of diabetic cataract consisted of anterior and posterior subcapsular clouds and dustlike opacities extending over the whole lens surface and tending to condense into greater complexes. In addition it is distinguished by opacities that bring out the superficial fiber design of the fibers, by the formation of small water-slits between the fibers, and by an extended subcapsular vacuolar degeneration.

The appearance of subcapsular fluid vacuoles as an early finding has been stressed by many other authors. It is assumed that the opacities develop from them.

Ephemeral Diabetic Refractive Changes. It is well known that with the onset of diabetes sudden changes in refraction may ensue. Either myopia or hypermetropia may appear suddenly in a person whose refraction formerly was emmetropic, or these changes may be added to an already existing error of refraction. Hence, a sudden unexplained change in refraction (especially myopia) in adults should be looked upon with suspicion. With treatment, the myopia may or may not disappear. When it does, as is usual, the individual may first pass over into a condition of hyperopia or if previously myopic, to a state of less myopia before returning to

* This term was applied to these opacities because of their resemblance to the figures employed in designating Roman numerals.

his original status. If the initial change is one of hyperopia (which statistically is more common), it tends to appear with decrease in glycosuria and in younger individuals* is accompanied frequently by systemic weakness. Without dwelling on the theories offered to explain the phenomena of refractive changes in diabetes (e.g., accommodative paresis and changes in refraction in the other ocular media) it should be pointed out that most authorities have attributed these changes to refractive alteration of the lens. Because of this, several writers have studied the lens during the course of a sudden change in refraction to ascertain whether there were any visible alterations biomicroscopically. The results were uncertain. I recently saw three cases, all women, aged 28, 32, and 54 respectively. In the younger two, the changes consisted of the sudden appearance of myopia and in the older one, hyperopia. The refractive change in the 28-year-old woman, who previously was emmetropic, appeared within 2 days and was the first known symptom of her diabetes. During these 2 days a gradually bilateral progressive myopia reaching three diopters developed. Insulin therapy was instituted 3 days later, and within a week the myopia began to recede. Two weeks following the onset of the myopia, the refraction was again normal. She did not pass over into hyperopia. In the second case, the interesting feature was that 1 diopter of myopia suddenly developed in the right eye and only $\frac{1}{4}$ diopter in the left. Three months previously under cycloplegic refraction, she accepted +0.50 cylinder axis 90 degrees in both eyes. Six months later, with the diabetes controlled by 15 units of protamine zinc insulin daily, there was no diminution in the myopia. In the third case—the eldest of these three persons—the patient suddenly noticed marked blurring in her distant vision as well as being unable any longer to read with her usual presbyopic correction of +2.25 sphere in both eyes. She now required a +3.50 sphere bilaterally for distance and +5.00 sphere for near vision. Three weeks following stabilization of her diabetes the hyperopia disappeared. About 6 months later she again showed a transitory increase in hyperopia associated with

* According to Granstrom⁴⁵⁸ this is rare over 30 years of age.

a return of a high degree of hyperglycemia. Her insulin dosage was increased markedly and within 10 days her hyperopia again abated. During the last year there have been no refractive changes.

I took special pains to examine these cases biomicroscopically at frequent intervals. In the first case, the anterior cortex (using the thickness of the corneal optic section for comparison) appeared thicker during the period of myopia than later. I was unable to convince myself of the presence of any definite blurring of the stripes of discontinuity or any increases of relucency in the cortex or nucleus.* In the other two cases I could not observe any biomicroscopic changes in the lenses either during the period of refractive change or afterward. In a case described by Vogt, a 44-year-old man, previously emmetropic, developed 3 to 4 diopters of myopia; a blurring of the bands of discontinuity (adult nuclear and fetal nuclear stripes) was observed. There was no increase of nuclear relucency as is the case in a lens with double focus. The chamber depth remained constant. With lessening of the myopia, the increased visibility of the zones of discontinuity become noticeable. At times Vogt had the impression that the sagittal thickness of the lens was increased. If the myopia results from increased difference of the index of refraction between the nucleus and the cortex, i.e., owing to decrease of cortical index or increase of nuclear index, then the zone of discontinuity should be more refractile. It could be hazarded that with the increase of cortical thickness, its action as a negative meniscus becomes less as compared to the positive action of the enclosed nucleus.

TETANY CATARACT (CATARACTA PARATHYROPRIVA)

The association of tetany and zonular cataracts has already been mentioned in the chapter on developmental cataract. This type of cataract is by no means pathognomonic when observed in cases of spontaneous tetany (Figs. 383, 384). However, it may well be

* Examination of this patient 18 months after the initial episode of the myopia revealed no refractive changes. However, biomicroscopically a single layer of delicate punctate glistening opacities at the level of the anterior and posterior lines of disjunction appeared. These seemed to develop peripherally and gradually faded out axially and evidently represent the first indications of the onset of a diabetic cataract.

that cataract of the zonular type is more characteristic for the cases in which tetany develops prenatally or in the first years of life, while those which occur later (whether they are spontaneous or follow operative damage or removal of the parathyroids) tend to be of the subcapsular type. In two recent cases in young persons called to my attention, the opacifications appeared as the typical complicated subcapsular variety similar to that seen following inadvertent parathyroid damage in thyroidectomy. The fact that cataract in spontaneous tetany may be similar to that in postoperative tetany (*tetania strumipriva*) was pointed out by Vogt who described the first biomicroscopic picture of tetany cataract in 1921. In the two spontaneous cases just mentioned, one occurred in a 12-year-old girl and the other in a 20-year-old woman, 3 months postpartum. In the latter case, treatment by Dr. Werner with dehydrotachysterol prevented tetanic attacks for the one and a half years of observation but no diminution in opacity of the lens was noted (Fig. 422 A, B, C). The vision remained unchanged at 20/80 in each eye until both lenses were removed by linear extraction. In the former case frequent tetanic attacks started just after the menarche. The vision was 20/50 in the right eye and 20/40 in the left. There were definite signs of previous hypoplasia of the enamel of the teeth and slight indications of past rickets. Therapy with dehydrotachysterol had just been started at the time of writing so that no definite conclusion concerning its effect on the progression of the opacities is as yet possible. It would be interesting to try this therapy in cases at the time of the earliest appearance of the lens opacities, especially in the light of Goldmann's experiments⁴⁵⁴ (discussed later). However, the difficulty in obtaining early spontaneous cases with lens opacities is great, since the condition of parathyropriva is not always recognized immediately.

The opacities in both cases were similar morphologically to those described in other endocrine cataracts (diabetes, myotonia atrophica). Nuclear cataracts and even total cataracts have been reported in tetany. According to both clinical and experimental evidence, it is known that the basis for tetany (both spontaneous and postoperative) is hypocalcemia. Experimentally in animals the

effects of parathyroidectomy on the production of tetany and cataract has been established. Goldmann showed that the tetanic convulsions and the rate of formation and regression of lens opacities (the appear-

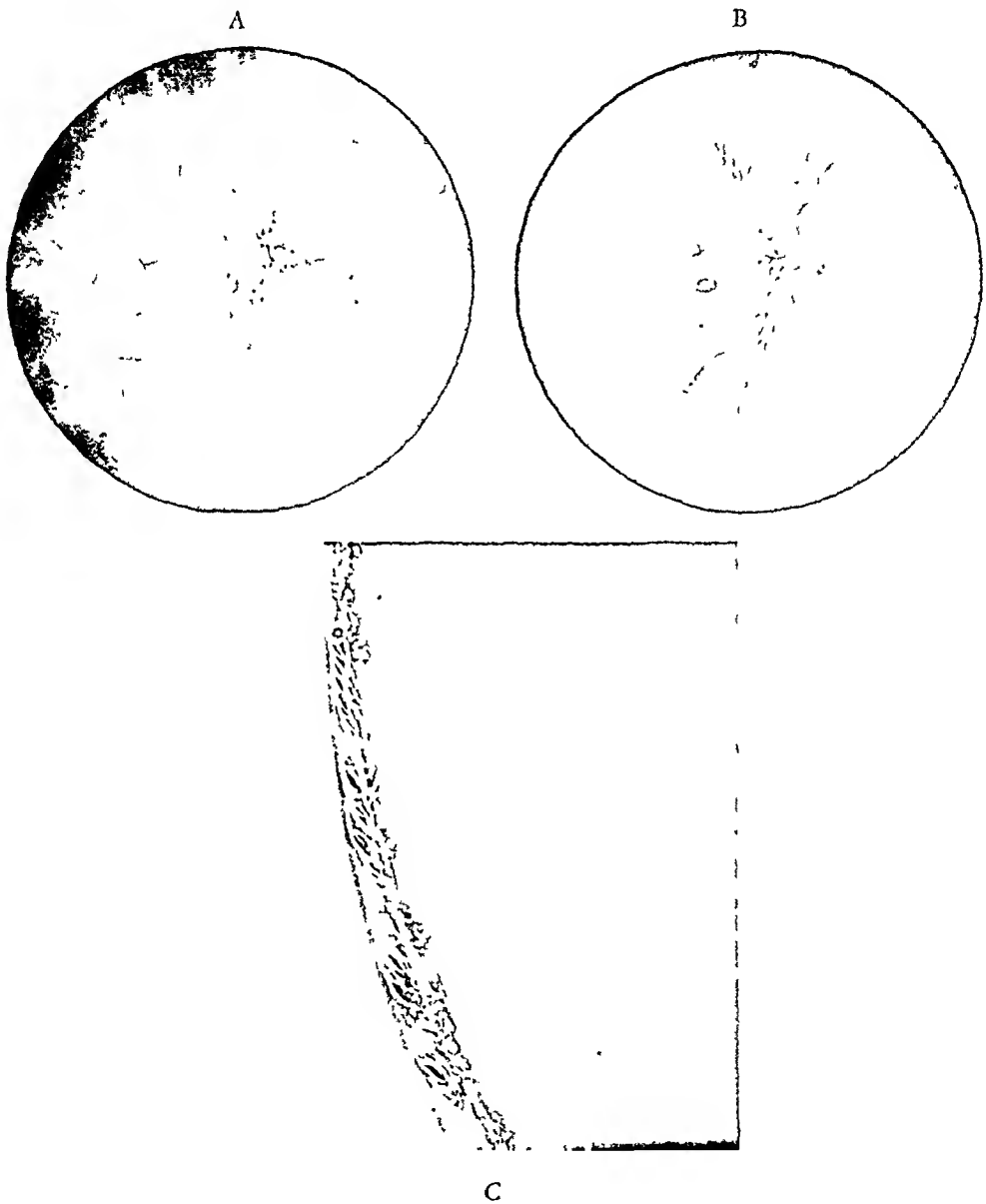


FIG. 422. Tetany cataract (*cataracta parathyropriva*). A. Right eye, diffuse view. B. Left eye, diffuse view. C. Direct focal illumination (optic section showing location of opacities).

ance of which is dependent on the attacks) could be controlled by the administration of calcium. Goldmann concluded that "cataract resulting from muscular tetany is not a chronic condition but a rapid, acute poisoning of the lens fibers. The toxic action is short-lived so that only the superficial layers are attacked. If the attack

is not repeated, lens turbidity may regress; if repeated, the fibers may be irreparably damaged and death supervene terminating in a total opacity of the affected area.” *

The typical location of the opacification of cataracta parathyropriva is subcapsular. This is, as described above, also found in juvenile diabetic cataract, cataract of myotonia atrophica, and cataract resulting from the action of exogenous toxins or physical agents. Consequently, the presence of this type of subcapsular opacification does not, of itself, permit an etiologic diagnosis. A thorough survey of the pertinent history and physical findings as well as laboratory studies must be instituted. However, Vogt stresses the point that the conspicuous appearance of the fiber design of the cortical surface (as sometimes occurs also in diabetes) is a typical finding and should cause one to be suspicious of tetany cataract. This would be particularly so in cases in which there is a history of a previous thyroidectomy. In the literature, there seems to be a wide divergence as to the time of onset of lens opacities following the operation, varying from a few months to years. The majority of cases developed subcapsular cataract, as long as 20 years following thyroidectomy. However, whether this was a result of the thyroidectomy is questionable, since the possibility of the appearance of coincidental presenile or senile cataract formation arises. Especially since it is known that in some cases the latter may begin subcapsularly and may resemble anterior cataracta complicata (e.g., anterior saucer-shaped or cupuliform cataract). Morphologically the opacities in tetany cataract may start with the presence of subcapsular dust-like dots or flakelike structures, with occasional iridescent crystalline formations. In the vacuolar type the development is similar to that described under the heading of anterior complicated cataract. In the latter type of tetany cataract, the fiber design is common, and the opacity itself is thin and tends to form irregular radiating bands in the direction of the suture system. With time, progression of the subcapsular opacities may result in complete opacification of the cortex, presenting a white pupil or “total” cataract.

* Quoted from Bellows.⁴⁰⁴

CATARACT OF MYOTONIA DYSTROPHIA (MYOTONIA ATROPHICA)

After Thomsen⁶³⁷ reported the symptoms of myotonia congenita (myotonia without atrophy) in 1876, Erb (1886)⁴²⁰ described atypical cases of myotonia with muscular atrophy. However, it was not until 1909, that both Batten and Gibb³⁵⁶ and Steinert⁶¹⁹ formulated the syndrome of myotonia atrophica as an entity characterized by atrophy of certain muscle groups associated with active or passive myotonia, i.e., a tonic spasm of certain voluntary muscles with slow relaxation. The classic example of myotonia atrophica is the inability to open the closed fist. Atrophy of muscle groups, e.g., facial, masticatory, and flexors of forearms and feet, etc., incapacitates these patients, the result of the first being masklike facies. Symptoms of this disease may become manifest just before or after puberty but in most instances they appear between the ages of 18 and 30 years. The average span of life is short. The condition is unquestionably hereditary, appearing as a dominant character. According to Allen and Barer,³⁴⁵ "Although the etiology is unknown, Fleischer has shown dystrophia myotonica to be a heredofamilial degenerative disease exhibiting anticipatory signs through several generations before it develops in its entirety in one generation. These anticipatory signs are: frequent instances of celibacy, many childless marriages, high infant mortality rate, and cataracts. In earlier generations cataracts develop at an earlier age, until in the 'myotonic generation' they are definitely presenile." Most authors consider that endocrine dysfunction is the basis of its pathogenesis.

In addition there is the tendency to loss of hair, disturbance of the salivary and sweat glands, loss of body weight, atrophy of the gonads, as indicated by early cessation of menses and testicular atrophy and impotence, and finally cataract. The appearance of cataract, first noted in this condition by Greenfield (1911),⁴⁶⁰ may be one of the earliest extramuscular symptoms and is so persistent and significant that its presence is now regarded as an important point in differential diagnosis. Earlier writers ascribed a lower percentage of cataract (10 to 30 per cent) to this condition. Such a low incidence may have resulted from the inclusion of allied neuro-

muscular disorders (e.g., myotonia congenita or amyotonia congenita), all of which do not exhibit cataract formation.

Recently I had the opportunity of examining biomicroscopically the eyes of 72 cases of muscular dystrophies collected by Dr. A. Milhorat from the Medical Service of the New York Hospital. The results are presented in the accompanying table.

TABLE XVIII
APPEARANCE OF CATARACTS IN CERTAIN
MUSCULAR DISEASES

DISEASE	NUMBER OF PATIENTS	LENS OPACI- TIES	TYPICAL SUBCAP- SULAR OPACI- TIES	CORO- NARY OPACI- TIES	CERU- LEAN OPACI- TIES
Myotonia trophica *	8	6	5	0	1
Myotonia congenita	4	0	0	0	0
Amyotonia congenita	2	0	0	0	0
Progressive muscular dystrophy	30	2	0	2	0
Myasthenia gravis	9	1	0	1	0
Periodic family paralysis	2	0	0	0	0
Dermatomyositis	4	0	0	0	0
Amyotrophic lateral sclerosis	6	1	0	0	1
Progressive peroneal sclerosis	10	1	0	0	1
Acute poliomyeli- tis	2	0	0	0	0
Total	72	11	5	4	2

* The two patients without opacities (of these one patient has a brother with no symptoms of myotonia atrophica but with typical lens opacities) had very mild symptoms, and the diagnosis of myotonia atrophica was doubtful. In the one patient with cerulean opacities, the case was unusual in that there was also peripheral neuritis.

A study of this table shows that the deep subcapsular variety of cataract is not only peculiar to myotonia atrophica but that it was found in practically every proved case. These findings are in accordance with those reported by Vogt (1922),⁶⁵¹ Waring, *et al.* (1940),⁶⁷² Allen and Barer (1940),³⁴⁵ Sautter (1941),⁵⁹⁷ and others.^{356, 429, 620}

The earlier descriptions of the lens changes in myotonia atrophica by Fleischer, Hauptman, and von Szily — as well as later by Allen and Barer — stressed the frequent appearance of a rosette in the posterior cortex.* With this they described the presence of senile white punctate opacities in both anterior and posterior cortical layers. Later in association with white punctate, angular or linear spots Vogt added another finding, i.e., smaller red or green iridescent dots, which were present in nearly all of his cases. The fact that in some cases there were well-formed crystals resembling cholesterol in the adult nucleus led to the supposition that the smaller colored dots (admixed with the larger white ones) were of the same substratum. However, an occasional colored dot may be seen in the cortex of normal lenses as well as in those having senile cataract or in tetany cataract. Vogt states that we should think of the possibility of myotonia when these colored dots are found in the anterior or posterior cortex in a greater number, particularly when they are associated with white angulated dots. The presence of all these opacities in the form of a dense layer, usually deep in the cortex, leaving the anterior line of disjunction and superficial cortical regions unaffected, tends to give the deeper parts of the lens a veiled appearance and often obscures the inner zones of discontinuity. In two of our cases there were a few isolated subcapsular vacuoles; the superficial fiber striping or design, so common in the cataracts of tetany and diabetes, is not characteristic of myotonia atrophica

* The presence of a rosette or starlike figure in the posterior cortex is by no means a universal finding in this condition. In the six cases which I recently examined, it was not found once, even in cases of long duration. Allen and Barer found that the earliest changes occurred posteriorly and consisted of minute white opacities and iridescent crystals and that they tended to concentrate along the sutures in a stellate manner. According to their findings the lens opacities seemed to progress in direct relationship to the rate of progression of the muscular symptoms. Cases showing rapidly developing muscular symptoms were accompanied by an earlier development of a "total" cataract. In only one of their cases did the opacities first appear in the anterior cortex.

(Fig. 423). With progression, which is usually slow, water-slits and lamellar separations develop, which in the end lead to maturity of the cataract. The axial concentration of the typical opacities and

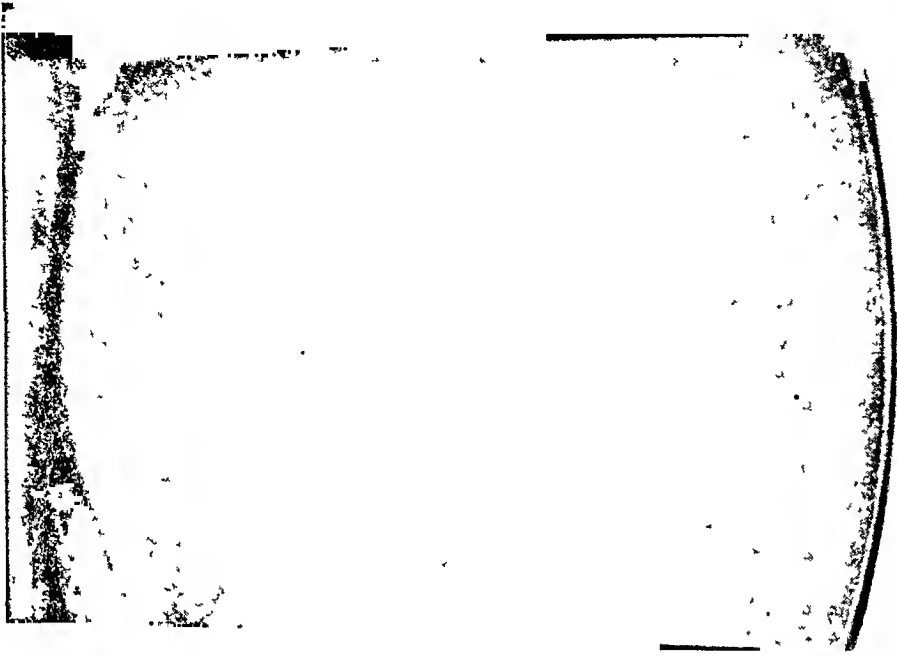


FIG. 423. Cataract in myotonia atrophica. Anterior and posterior subcapsular punctate opacities.

iridescent dots should differentiate myotonia cataract from coronary and cerulean cataract, although the later may often be present fortuitously, considering that its incidence has been computed as high as 25 per cent in otherwise normal individuals.

CATARACT ASSOCIATED WITH MONGOLIAN IDIOCY

The high incidence of cataract in Mongolian idiocy has been recorded by numerous observers. Bellows, compiling the statistical evidence of both Ormond (1911-1912)⁵⁵⁷ and van der Scheer,⁵⁹⁸ points out that cataract occurred 61 times in 102 of their cases (59.8 per cent). They found cataract only once in a person under the age of 8 years.* According to Koby who published the first bio-

* Although this might suggest that the cataractous changes are acquired rather than of congenital origin, it does not entirely preclude a hereditary tendency, in the same sense as in other opacities appearing during childhood or early youth (cerulean or coronary). However, in the present state of our knowledge dogmatism concerning the etiology of juvenile cataract is not justified. As previously mentioned, the recent reports on the finding of

microscopic description of cataract in Mongolian idiocy in a 17-year-old girl, lens opacities rarely appear before the fifteenth year. In his case the bands of discontinuity were fairly clear. There were

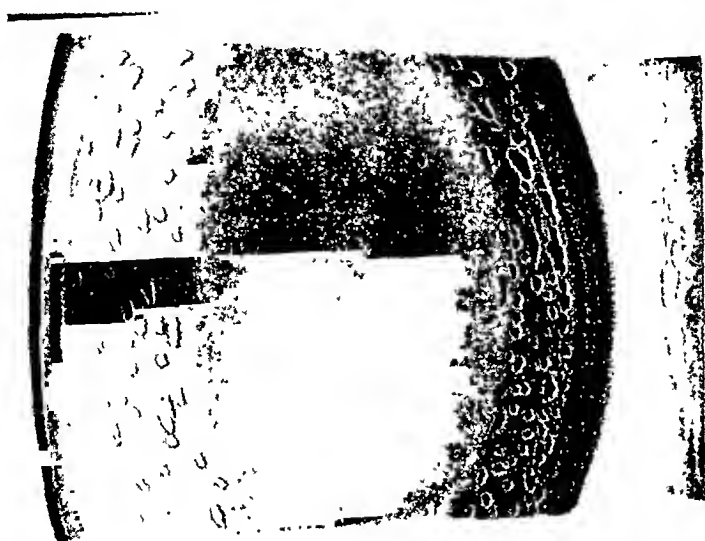


FIG. 424. Cataract in Mongolian idiocy. (After Koby.)

two types of multiple opacities: the flattened type, which were yellow and blue in color, similar to those in cerulean cataract, and the fine crystalline type showing green and blue iridescence not unlike those of cataract parathyropriva and myotonia atrophica. The opacities occupied the deeper cortex and parts of the adult nucleus, leaving the embryonal nuclei unaffected. From the descriptions in the literature, the morphologic appearance of cataract seen in Mongolian idiocy varies from case to case. This variance may depend on the fact that, having progressive tendencies, the appearance and localization of the opacities changes with age. The distribution and the typical punctate to flocculent character of the opacities may be suggestive of endocrine dysfunction as the etiologic factor (Fig. 424). As a rule, the opacities are denser axially, while in a more scattered fashion they may occasionally extend peripherally around the visible equatorial region. Ormond⁵⁵⁷ published illustrations in which the opacities formed a ring leaving the axial region free. Fre-

cataract in the newborn whose mothers had German measles during their pregnancy indicates the possibility of exogenous influence. Likewise the effect of endocrine dysfunction, or of nutritional and dietary deficiencies on production of lens opacities, especially in the young, still requires clarification.

quently they are more numerous in the posterior cortex than in the anterior, and they assume a radiating disposition. Duke-Elder in describing the opacities stated: "Some are powdery and punctate, some annular and some flaky; while others have a crystalline appearance. The majority are white, while others glitter with red or green light."

In addition to these cortical opacities, essentially similar to those found in myotonia dystrophica, van der Scheer⁵⁹⁸ has called attention to the finding of suture cataract (fetal [especially the anterior Y] and at times cortical) as an essential part of the lens changes in Mongolian idiocy. This change was not found in most of the cases cited in the literature by other authors. However, the frequency of occurrence of suture cataract in normal individuals should be considered. Hence suture opacification is probably not an essential feature of cataract associated with Mongolian idiocy but, when present, is probably coincidental.

CATARACT ASSOCIATED WITH CERTAIN SKIN DISEASES (CATARACTA DERMATOGENES)

The complication of presenile cataract in certain chronic diseases of the skin has been pointed out by numerous observers since the original observations of Rothmund (1869).⁵⁸⁵ Most of the cases described occurred with neurodermatitis. Less frequently they have been reported in scleroderma (as part of Werner's syndrome) or in polikiloderma atrophicans vascular (Rothmund's disease) and eczema. It is not surprising that in certain conditions in which the ectodermal tissues are primarily affected (skin, hair, nails, teeth, etc.) that the lens participates. At the present time there is no unanimity of opinion regarding the etiology of these skin diseases in which cataract may sometimes appear. In all these syndromes, dysfunction of the endocrine glands has been suggested but the evidence is inconclusive. Allergy has been advanced as a basis of the pathogenesis especially in the case of neurodermatitis by Daniel,³⁹¹ Brunsting,³⁷² and Beetham.³⁶³ Both Werner's and Rothmund's disease have a familial tendency, the latter being transmitted as a recessive char-

acteristic. The descriptions of dermatogenous cataract, in the literature, vary considerably. However, while there are certain characteristics that seem to be common to them, they are in a broad sense

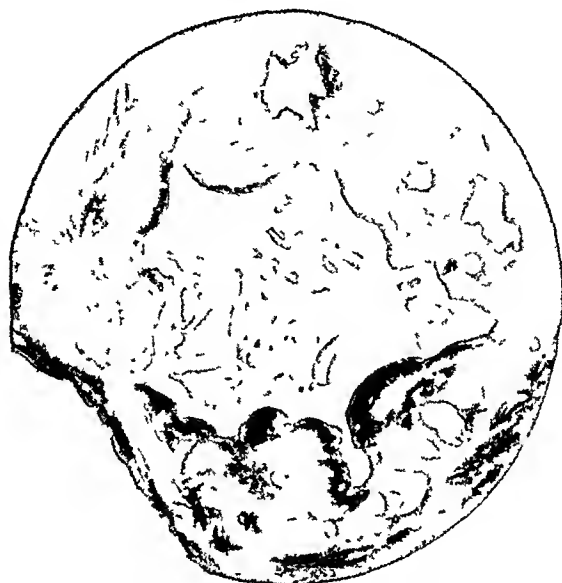


FIG. 425. Cataracta dermatogenes (After Vogt)

common to all the opacities found in endocrine dyscrasias (Fig. 425). These common characteristics are as follows: the opacities in the beginning are subcapsular and frequently follow the radiating design of the sutures; others tend to appear in the form of rosettes. In some cases the development of a central capsular opacity was noted. Vogt described a case occurring in a 28-year-old man, who since early youth had neurodermatitis. He had a milky lens with a denser irregular capsular opacity (resembling a plaque or shield) in the axial region. The plaque measured 3 mm. vertically and 3.5 mm. horizontally, and this practically filled the pupillary area. The edges of this capsular opacity were scalloped in an uneven concave manner and showed shagreen-free areas, demonstrating a change of level in the capsular curvature. In the right eye there was also an incipient anterior complicated cataract in the form of a rosette; the radiating opacities were subcapsular and in places a fiber design was apparent. The posterior cortex in both eyes at this time seemed unaffected. The vitreous structure was disturbed and contained numerous brown pigment granules. Three months later examina-

tion of the right eye showed an oval capsular opacity not unlike that in the left eye in which gaps in the shagreen reflex were found at the margins. The posterior cortex in this eye now showed a cloud of opacities axially. The change in curvature of the anterior capsule at the border of the capsular opacity can be demonstrated by the narrow beam where an actual depression or concavity results. This type of capsular alteration can occur also in hypermature cataracts both in the senile variety as well as in the complicated form. Both Lowenstein⁵²⁸ and Andogsky³⁴⁸ described similar forms of cataract in neurodermatitis. Andogsky described four cases in young persons in whom the cataract began as an anterior subcapsular rosette opacity. Recently I saw and operated on a case of precocious cataract in a young man, aged 28 years, who had marked neurodermatitis of the face and extremities. The lens opacities were bilateral and consisted of dense subcapsular radiating opacities, anterior and posterior. There were numerous subcapsular vacuoles. No axial capsular opacity was found, but there were several small flat ones distributed more peripherally in an uneven way. In the right eye the vision was reduced to 20/100, and this could not be improved. The vision in the left eye was 20/40+ with a -0.75 sphere. He was under observation for three months prior to the time cataract extraction was performed. During this time there was no change in visual acuity. In a case of Werner's disease (scleroderma),* reported by Agatston and Gartner,³⁴⁴ occurring in a 38-year-old man, whom I had observed for six years and on whom I had performed a cataract extraction in the left eye in 1937, there were typical findings of dermatogenous cataract. When first seen, this patient had anterior and posterior delicate subcapsular opacities in the axial regions of both lenses, more marked in the left eye (Fig. 426). There were numerous subcapsular vacuoles anteriorly. The opacities radiated in a manner suggestive of a rosette. In the periphery of both lenses

* At 32 years of age this patient already had the appearance of an old man. Partially bald, his hair was gray and the skin over the face and extremities was inelastic, thickened, and brownish in color. He was slight in build with small hands and feet. His teeth were carious. The configuration of the pubis and the arrangement of the pubic hairs were feminine. The penis and scrotum were small, all indicating a possible endocrine disorder.

there were scattered coronary opacities. At this time bilateral iridec-tomies were performed. There was little progression of the opacities or loss of vision during the next five years. In the sixth year the

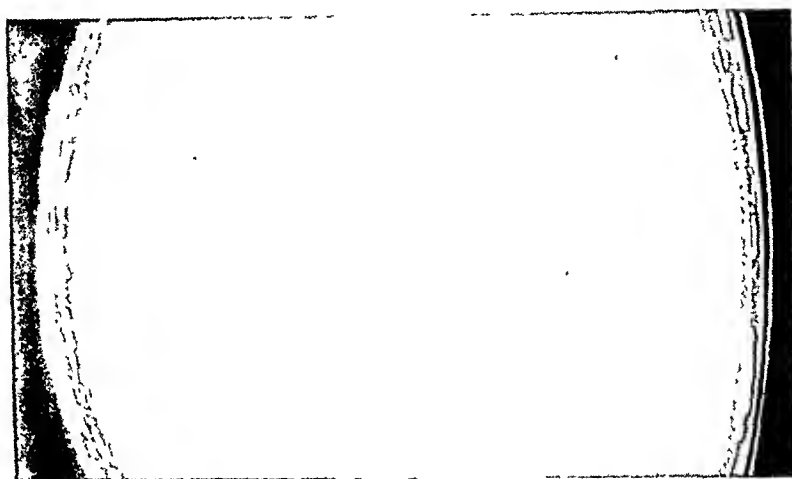


FIG. 426. Cataracta dermatogenes. Early changes. (Case of Agatston and Gartner.)

vision in the left eye began to fail, becoming reduced from 20/50 to 20/200. With the biomicroscope, the disintegration of the cortex was seen. Water-slits with spokelike opacities formed. In the posterior cortex there was a saucer-like opacity. The lens was extracted extracapsularly. The right eye was unchanged, vision being 20/40 unimproved. About a year later the cataract matured in the right eye and was removed by Dr. S. A. Agatston. According to the report of Agatston and Gartner: "Both eyes had operative colobomas of the iris above. The right eye had a cataract which matured in a few months. When the patient was first seen he had posterior cortical opacities, some subcapsular vacuoles and anterior peripheral cortical opacities."

TOXIC CATARACTS

Since the original communication of Bouchard (1886)³⁶⁹ in which he showed that cataracts could be produced in rabbits by feeding them naphthalene (1 gram per kilogram of body weight) mixed with their daily diet, numerous investigators have corroborated experimentally as well as clinically that opacification of the lens may follow in the wake of toxic effects resulting from the in-

gestion or inhalation of certain poisonous substances. Especially prominent among these poisons are ergot and thallium and benzene and phenol derivatives such as phenol, naphthalene, dinitrophenol, dinitro-orthocresol, and paradichlorobenzene. Also experimentally in rats, lactose and galactose feedings have produced lenticular opacification. Bellows summarized the findings as follows: "The susceptibility to cataract formation following poisoning by these substances shows marked differences in the various species. Poisoning by dinitrophenol or ergot produces cataract in man but has never produced cataract in lower animals; naphthalene* poisoning produces cataract in rabbits (Bouchard and Charrin) and in man (Lezenius⁵²¹; thallium induces cataract in rats but not in man; and thus far galactose cataract has been reported in rats only. Not only do the species differ in their response to these various poisons, but there is a marked individual susceptibility within certain species." I found a similar selectivity in the case of paradichlorobenzene poisoning in which cataract followed the inhalation of the vapor from this substance in humans and not in animals, although in both there were manifestations of toxic hepatitis. In humans, in most cases the appearance of toxic cataract (dinitrophenol, naphthalene — in one case reported by Lezenius, paradichlorobenzene and ergot) was characterized by a rather long latent period, i.e., the lens changes ordinarily do not appear immediately after the poisoning. In most cases the latent period extended over a year. For example, in an epidemic following the ingestion of rye harboring an ergot-containing fungus, Tepkjilaschin⁶²⁸ noted that cataract did not occur until from 2 to 5 years after the poisoning (27 cases). Likewise in dinitrophenol poisoning Rodin (2 cases)⁵⁷⁹ noted that the average latent period was about 15 months. In the two cases of cataract that I described following the inhalation of fumes of paradichlorobenzene, the incidence of the lens changes occurred 12 months

* As a result of the work of Bourne, Young and Stekol (cited by Bellows) the mechanism of naphthalene cataract formation may be reconstructed as follows: Naphthalene is detoxified in the body by conjugation with cysteine to form a mercapturic acid derivative. All tissue, including the lens, suffer a reduction in cysteine proportional to the amount of drug ingested. With the loss of cysteine the respiration of the lens (which is an avascular tissue) must suffer and if sufficiently interrupted, the lens must die with resulting opacification.

(bilateral) and about 7 months (uniocular) respectively after the patients were removed from the noxious fumes. The prolonged latent period and the rapidity with which the toxic cataracts develop after once having started have not yet been explained.

In the case of dinitrophenol poisoning there is some available evidence that *in vitro* the increase in metabolic rate and consequent depletion of the available oxygen may be a factor in the production of this form of toxic cataract. Because of the hepatic damage associated with toxic cataract, Berliner,³⁶⁷ following Onfray and Dreyfus,⁵⁵⁶ suggested several ways in which hepatic damage might be related to cataract formation. However, conclusive evidence of this relationship is still lacking.

The lens opacities in toxic cataract tend to start subcapsularly in forms not unlike those seen in endocrine cataract, viz., with punctate and vacuolar changes either discrete or in the form of rosettes and also with iridescent crystals. However, unlike the slower development ordinarily seen in complicated cataract, in most cases the process in toxic cataract is characterized by a more rapid progression. This is evidenced by quick cortical swelling and disintegration. Large water-slits and laminary separations are soon followed by complete opacification or maturity of the cataract. In one of the cases of paradichlorbenzene poisoning, mentioned above, within a week the vision failed from 20/20 to the perception of fingers at 3 feet. The anterior chamber became shallow, and intraocular pressure increased so that extraction became urgently necessary. When first seen, 3 or 4 days following the onset of sudden visual blurring, the vision in the right eye was reduced to 20/70. At this time there already was considerable swelling of the cortex. There was a subcapsular layer of fine punctate opacities and vacuoles. Beneath this in the anterior cortex, which was hazy, there were many large water-clefts, and in the periphery, areas of laminary separation. The nucleus was faintly relucant. Through the haze a slightly yellowish granular opacification, which in specular reflection revealed several glossy iridescent dots, could be just made out subcapsularly in the posterior cortex, reminiscent of posterior complicated cata-

ract. Within a few days no further details could be discerned in the nucleus or posterior parts of the lens owing to the total disintegration of the anterior cortex (Fig. 403 A, B). The condition in the left eye followed the same rapid course. In a case of ergot poisoning, Cattaneo³⁸³ reported the findings of anterior and posterior sub-capsular rosette-like formations. Whalman,⁶⁷⁶ who tabulated the findings in 27 cases of dinitrophenol cataract, stressed this rapid development and bilaterality and summarized their clinical appearance as follows: "Study of these cataracts with the biomicroscope revealed certain characteristic changes which appeared in all instances. There is no abnormality in the cornea. The aqueous humor shows an increased flare. The anterior capsule of the lens is at first spotty, dry and lusterless, and later somewhat pebbly, while beneath the capsule fine gray cloudy opacities appear. Still later irregular pearl-gray opacities appear in the deeper layers of the cortex. Concurrent with the early changes in the anterior portion of the lens, a polychromatic luster can be seen in the zone of specular reflection in the posterior cortex. These alterations are followed by a marked swelling of the lens, and the embryonic suture lines seem to be completely shattered by dark spaces resembling water clefts. If the condition is seen later than this there is almost complete disorganization of the lens and nucleus." In one case — that of a 48-year-old woman — I found bilateral nuclear cataracts with incipient cortical changes. The blurring of vision began 6 months subsequent to a course of "reducing" therapy in which thyroid extract was alternated with the administration of dinitrophenol. During a period of 6 weeks in which she lost about 22 lbs. of body weight she took approximately 15 gm. of desiccated thyroid and about 20 gm. of dinitrophenol. Disregarding this medication and sudden onset of the lens opacification, morphologically this cataract could easily have been mistaken for an ordinary senile cataract.

In an analysis of the reported cases, the amount of the dinitrophenol or the length of time which it was administered seemed to bear no definite relationship to the onset or progression of the cataracts. In one of Whalman's cases cataracts appears 3 months

following the daily ingestion of 300 mgm. during one month only. Three weeks after their onset they became mature.

CATARACT ASSOCIATED WITH GLAUCOMA

The association of cataract with glaucoma has been known for a long time, particularly the form that occurs in absolute glaucoma (in blind eyes where, owing to iris degeneration, the pupil is wide) and it is characterized by a distinctive porcelain white, yellow, or greenish hue.* In 1879 Priestley Smith⁶¹¹ described this type of cataracta glaucomatosa, which usually starts with nuclear opacification and is frequently accompanied or followed by posterior saucer opacities and cataracta complicata. Gradually the cataract becomes total, eventually passing over to the hypermature state in which shrinkage occurs (as evidenced by capsular folding, and deepening of the anterior chamber). At times following zonular stretching or rupture, dislocation results. Secondary chalky and crystalline degenerative changes are often manifested. In some cases, especially when the lesion is extensive, iridic adhesions are present, disintegration and absorption of lens material may lead finally to a condition in which only a folded membrane is seen in the pupillary area.

Besides this form, biomicroscopic examination of the lens in glaucoma, especially in chronic cases in which surgery has been resorted to, will almost invariably reveal lens changes. These may vary from small capsular and subcapsular opacities, frequently localized to the segment corresponding to the operative procedure (iridectomy, iridocleisis, cyclodialysis, or trephine) to more extensive cortical or nuclear cataract. The rapidity with which these opacities follow surgery would suggest that they are in all probability directly or indirectly related to it. As in traumatic cataract, localized subcapsular opacities may form; and when rapid swelling of the cortex ensues, a mature total cataract results. In the latter instance rupture of the lens capsule may occur either as a result

* Before glaucoma was known as an entity a distinction was made between gray and green cataract. The latter was deemed inoperable.

of instrumentation (particularly when the anterior chamber is shallow) or spontaneously (the posterior capsule being especially vulnerable) following sudden decrease in intra-ocular pressure

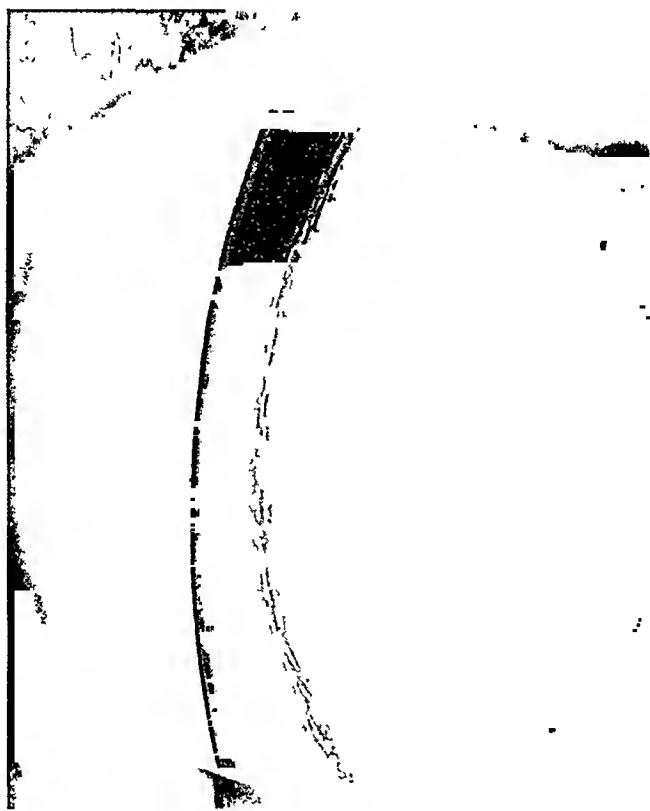


FIG. 427. Disseminated cataract after glaucoma. A layer of opacities seen three months following iridectomy (not present before operation). Cortex wider than normal. Beginning nuclear opacification.

on opening of the anterior chamber. A variety of other causes could account for the appearance of a rapid cortical swelling and disintegration, even if it were possible to rule out actual rupture of the capsule, e.g., increased capsular permeability, or the action of toxins or other factors initiated within the lens itself (Fig. 427).

Under the designation "*cataracta disseminata subepithelialis glaucomatosa acuta*" or "*white glaucoma spots of the lens*," Vogt (1930)⁶⁶⁰ described the sudden appearance of multiple sharply circumscribed white subcapsular opacities seen only after an attack of acute glaucoma. At first he thought that these opacities were conditioned by the operative procedure (following the sudden reduction in intra-ocular pressure), since they were seen after iridectomy.

But later, he found the same type of spots in unoperated cases of acute glaucoma that were controlled by miotics. In optic section the thickness of the opacities is uniform and, being thin, they tend to lie

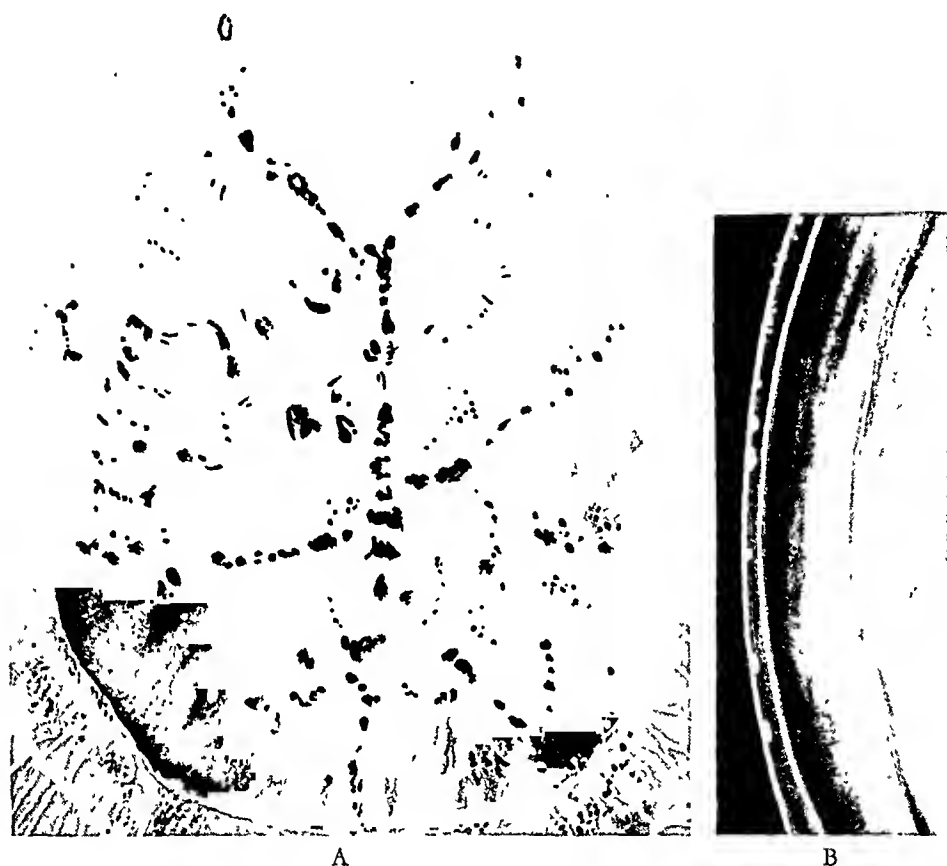


FIG. 428. Disseminated subepithelial cortical cataract of acute glaucoma. A. Diffuse illumination. B. Optic section. (After Vogt.)

in one plane parallel to the surface. They vary in size (from 0.3 to 0.02 mm. or less) and in shape are principally roundish or angular. Predominantly these spots are found in the axial region, where they string out in the direction of the suture lines (Fig. 428 A, B). According to Vogt their surface has a smoothness, reminding one of polished ivory. Characteristically the spots, which appear within days or weeks following an attack of acute glaucoma, are subcapsular, the shagreen of the capsule over them not being interrupted. This together with the radial direction of their extension (in one plane following the direction of the cortical sutures) suggests that they are

not related to the anterior capsule in the sense of deposited exudative spots or to the denudation of the underlying epithelium. Within short periods of time (weeks to months) the narrow beam will disclose that these spots tend to be displaced deeper into the cortex. As in polar cataract and in certain types of traumatic opacities,* the ingrowth of new lens fibers subcapsularly serves to push these opacities back so that with time a lucid interval of increasing width is observed between them and the anterior line of disjunction. In one case the subepithelial opacities, which were noted immediately following an acute attack of glaucoma, after five years were seen to be located in the neighborhood of the adult nuclear surface. The opacities do not seem to enlarge with time but in the main either remain stationary or in some instance fade or become smaller. In another case described by Vogt, typical subcapsular opacities were seen in a case of acute glaucoma one week post-operatively. The following day it was observed that they were already smaller in size. This might indicate that they are capable of being absorbed when fresh and when located in their original subcapsular location. Later the possibility of partial absorption seems to be less, especially after the opacities have pushed into the depth of the cortex by the ingrowth of newly formed healthy fibers. In all instances, the posterior cortex never showed any similar foci or other characteristic findings. The fact that these spots have not been noted more frequently in cases of acute glaucoma that were treated by miotics might be explained by the suggestion that once the attack is over and the pressure is controlled it is rare for a physician to hazard dilatation of the pupil for diagnostic purposes only.

Considering the uniformity of the picture in which small intensely white subcapsular flat opacities are seen following attacks of acute glaucoma Vogt considers *cataracta disseminata subepithelialis anterioris* to be a distinct and separate entity. In view of the acute onset of these opacities — viz., following an attack of acute

* Similar opacities have been noted following severe contusions. In these cases an increase in intra-ocular pressure (although of shorter duration) may be a factor in the genesis of the opacities.

glaucoma — one might not be incorrect in calling this entity an example of "acute cataract."



FIG. 429 Deformation of the lens by a tumor.

DEFORMATIONS OF THE LENS BY LOCAL PRESSURE FROM TUMORS OF THE CILIARY BODY

Tumors (especially melanosarcomas or leukosarcomas) of the ciliary body extending in the direction of the lens may by local pressure cause deformation and indentation of the lens at the point of contact. Such an indentation may result in a compression of the outer softer cortical substance so that the distance between the outer bands of discontinuity (lines of disjunction and adult nuclear band) may be narrowed (Fig. 429). In spite of the deformity no opacification in the compressed area may result. In one case of this type Vogt found a large water slit surrounded by vacuolar masses in the region of the lens between the posterior capsule and the posterior line of disjunction. Here, the local pressure resulted in a concave distortion of curvature within the lens so that the posterior band of disjunction was pushed toward the surface of the adult nucleus.⁵⁵²

SPONTANEOUS DISLOCATION OF THE LENS

Generally, spontaneous or nontraumatic dislocation of the lens follows some pre-existing ocular disease. Weakening (stretching)

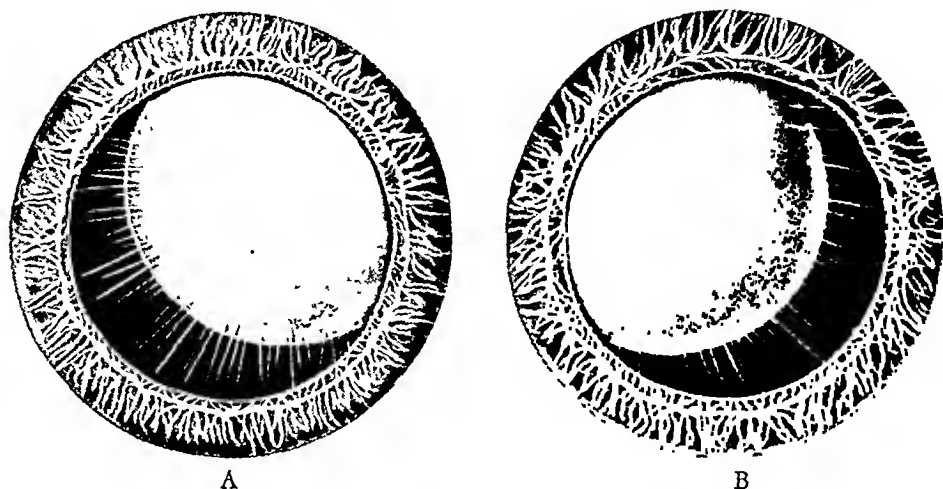


FIG. 430. Dislocated lens (Marfans syndrome). A. Right eye. B. Left eye. (After Vogt.)

or disintegration of the zonule results in either a partial (subluxation) or complete (luxation) displacement of the lens. In the latter case (like a free balloon) it becomes completely detached from all its zonular and vitreous attachments and no longer occupies the fossa. In this case owing to the effects of gravity, it tends to sink. Depending on the fluidity of the vitreous and other local factors, the luxated lens may be confined to the vitreous, at times floating freely within it or depending on the size, width of the pupil, length of the zonular fibers, restraint of the vitreous and position of the head, it may wander back and forth from the vitreous to the anterior chamber. A sudden strain — from sneezing, coughing, or bending — may precipitate such displacement. Traction by inflammatory membranes may also lead to luxation of the lens. With tumors, and in cases of absolute glaucoma where the pupil is wide, a progressive subluxation of the lens may lead to complete luxation. Following luxation into the anterior chamber, the lens may at first remain clear, appearing like an "oil drop," but as a rule it soon becomes opaque. Opacification of the cornea (edema, folds, etc.)

usually ensues. Cases have been reported in which ulceration of the cornea and extrusion of the lens occurred.

However, with subluxation in which the separation is incomplete, the unaffected parts of the zonule tend to pull the lens in a direction opposite to location of the torn or stretched zonular fibers. As a consequence of the eccentric position of the lens aphakia may result, provided the lens is sufficiently displaced from the pupillary area (Fig. 430). This may lead to a monocular diplopia where the vision in the affected eye occurs partly through the phakic portion of the pupil and partly through the aphakic portion. In subluxation the lens may be tilted forward, backward, or laterally and consequently may cause variation in depth of the anterior chamber, an effect that is easily recognizable with the biomicroscope. In most instances the free edge tilts toward the vitreous.

Disregarding the hereditary and congenital forms of lens displacement (e.g., ectopia lentis simplex — uncomplicated by other deformities — or complicated by other defects, especially Marfan's syndrome) spontaneous displacements are found, chiefly in high myopia, hydrophthalmia, hypermature cataract, and uveitis.* In all of these the necessary requisites, i.e., zonular and vitreous degeneration, for freeing of the lens are present (Plate LXXVIII). In hydrophthalmos, owing to distention of the globe, a breakdown or disintegration of the zonule may result in eventual luxation of the lens. In certain instances a breakdown of the capsule may occur and the contents of the lens may be extruded, the capsule remaining *in situ*. Jess saw such a case in the hydrophthalmic eye of a cat.

In contradistinction to the hereditary forms in which the subluxation is usually stationary, deterioration in the zonule and vitreous in cases caused by disease tends to be progressive so that ultimately total luxation of the lens may ensue.

From the standpoint of biomicroscopy, displacement of the lens produces a striking picture. As the focused beam passes through a subluxated lens in the vicinity of the displaced side (dilated pupil),

* Since the hereditary forms of dislocation of the lens are associated with defects in the zonule, they will be considered in the chapter dealing with the zonule.

internal scattering of the light produces a bright reflex that outlines the equatorial rim of the lens (Fig. 459 A, p. 1344). In contrast to the bluish-gray relucency of the lens, the aphakic portion of the pupil appears jet black. In this aphakic area the focal beam reveals the zonular fibers which may either be stretched or partially missing, but it is rare not to find some evidence of them. Frequently broken fragments of the zonule will be seen hanging from the capsule near the lens equator (Plate LXXVIII). At times such fibers may be connected to a completely detached zonular lamella, which appears like a veil separated from the capsule. Several authors have reported cases in which the lens was luxated out of the enveloping zonular lamella (Jess,⁴⁸⁹ Meesmann⁵⁴¹ and Stein⁶¹⁷). The tendency to opacification is greater in the luxated lens than in one that is only subluxated. But in either case an accompanying inflammation or increase in intra-ocular pressure will predispose to lenticular opacification, which is manifested as posterior complicated cataract, or subcapsular vacuolization commonly. In addition a form of zonular opacity that, like *cataracta coronaris*, outlines (in optic section) the curve of the adult nucleus at its equator may be found. The tendency for the opacification to become total is not unusual. In subluxation in spite of the apparent deficiency of zonular tension the biomicroscope will usually show the normal peripheral divergence of the anterior line of disjunction. This perhaps might indicate that the tension of the zonule only plays a part in the production of the phenomenon and that, as mentioned before, the major part of it results directly from the relative thinning and widening of the fibers axially where they abut the branching sutures.

CERTAIN PATHOLOGIC ALTERATIONS OF THE LENS CAPSULE

Considering the importance of the lens capsule, very little is known concerning its origin, morphology,* physical and chemical

* The fact that the hyaline or cuticular capsule (probably formed as a secretion of the lens epithelium) was composed of numerous lamellae was established by earlier writers, such as Kolliker,⁵⁰⁸ Ivanow and Arnold,⁴⁸⁷ Berger³⁶⁵ and Schirmer⁶⁰⁰ and others. By maceration, Vogt found ten layers; the superficial (oldest) being the thickest. The deeper (towards the epithelium) gradually becoming thinner.

functions or its exact role in the mechanism of accommodation. Likewise little is known about its physical properties, e.g., variations in elasticity during life. From histologic preparations it is known that the capsule thickens with age. This also is seen in cataract (especially in the hypermature and complicated forms).

Lately our attention has been called to the importance of the capsule in the regulation and maintenance of lens metabolism. Considerable experimental work has been done on capsular permeability, in an effort to explain the exact role of this function in health and in disease. As it now stands we are confronted by a mass of controversial physical and chemical evidence that must await future clarification. Further, our interest in the capsulo-zonular diaphragm has increased with the modern trend toward intracapsular cataract surgery.

Clinically with the aid of the biomicroscope, the finding of certain other capsular changes (e.g., folds and exfoliation of the superficial lamellae) has substantiated many of the histologic observations. According to Vogt, four important clinical facts have been ascertained biomicroscopically: (1) Folding of the capsule—the formation of capsular folds is an early indication of shrinkage of the lens. This is seen in hypermature cataract and as a symptom of lens injury or traction by scars. The normal capsule produces fine delicate folds, those of the thickened capsule are coarse and sausage-like. (2) The normal capsule is always smooth, there being no such condition as a physiologic depression. An exception to this occurs in the presence of total sclerosis where localized irregular unevennesses in the capsule may occasionally be found. (3) The discovery of the detachment of the superficial layer of the capsule in glassblowers or those exposed to the action of heat (infrared cataract). (4) The so-called “senile” exfoliation of the superficial capsular lamellae, which may be attended by glaucoma (glaucoma capsulare).

Under this heading are included anterior and posterior capsular changes, principally associated with inflammation and charac-

terized by the deposition of exudates and pigment and by the presence of vessels and linear structures (pleats). Genuine capsular folds are discussed on page 1140. It goes without saying that the above-mentioned capsular changes are also found secondary to inflammation induced by injury or operation. For the proper study of these capsular alterations dilatation of the pupil is usually necessary. In diffuse illumination, subcapsular changes, e.g., anterior complicated cataract (page 1161), might conceivably be confused with purely capsular alterations. With the focal beam, especially the optic section, the capsule stripe will become visible as a line and the above error can be avoided easily. Changes peculiar to the hyaline capsule will cause a change in direction (elevation or depression) of the first line of discontinuity (the capsule stripe).

Other capsular alterations, such as the special changes (capsular and subcapsular) peculiar to the action of heat (infra-red), to contusions and perforating injuries (e.g., capsule ruptures, Vossius' rings, siderosis and chalcosis, etc.) are considered elsewhere (Chapters 27 and 28). The so-called "senile" exfoliation of the anterior lens capsule is also included in the chapter on pathologic changes of the capsule, rather than under the heading of a senile lens change. In spite of the fact that it rarely occurs before the sixth or seventh decade, its relationship to glaucoma and cataract would seem to warrant that it be considered as a pathologic entity rather than a purely senile physiologic change.

INFLAMMATORY DEPOSITS ON THE ANTERIOR LENS CAPSULE

Biomicroscopically, during or in the wake of intra-ocular inflammation, depositions on the anterior capsule in the form of exudates or pigment are a commonly observed occurrence (Fig. 431). In some instances, e.g., pigment stars and gray threads, it may at times be difficult from the standpoint of their morphology alone to distinguish between those resulting from inflammation and the physiologic rests of the tunica vasculosa lentis. As a rule in doubtful cases, careful consideration of the history and minute examination of the



A



B



C



D

FIG. 431. Various types of inflammatory capsular deposits. A. Starlike pigment deposits on anterior lens capsule. Note synchia where iris stroma is drawn over pupillary margin. B. Membranous and fibrillar exudate on anterior lens capsule. C. Exudative membranes over anterior lens capsule. Note pleatlike folds. D. Exudative deposits on anterior lens capsule arranged in radiating manner.

cornea, iris, and vitreous of both eyes for evidences of inflammation assist in making a differential diagnosis. However, it must be admitted that in cases of mild or ephemeral iritis, especially in the



FIG. 432. Inflammatory deposits on the anterior lens capsule.

absence of signs of synechiae, such a differentiation may be exceedingly difficult.

Following iritis, the well-known deposits of irregular reddish-brown pigment occur on the capsule in the isolated form or in clumps or as a broken segmented ring outlining the place of contact of the pupillary edge and capsule; these become more evident on dilatation. When large they may be seen with the unaided eye. There may be fine pigment dust only, or large clumps may be surrounded by pigment dust. In addition to large clumps and amorphous grains, smaller circles of pigment may be found rarely occurring in cases of quiet iritis. With high power the surface of the larger pigment clumps will be observed frequently to have a finely granulated appearance. Also the deposits may be surrounded in places by a delicate grayish exudate, which possibly acts like a cement substance. At the site of a synechia such pigment masses may become stretched out, owing to pupillary action (see chapter on iritis). (Plate LXXIII.)

Exudate on the anterior lens capsule may assume various shapes and forms, the appearance of which frequently changes with time. When early and near the pupillary margin, it may resemble little dots, balls (efflorescences), or flat veils (Fig. 432). These can give rise to twisted threads which, when crossing one another, resemble loosely meshed nets or starlike figures. Upon these nets starlike

figures may be distributed. In some cases the pigmentation may be heavier and thus may hide the delicate grayish threads, so that only a layer of branching pigmented stars is seen. Pigment stars and threads may be arranged in irregular whorls or may extend themselves like a wreath (similar to Vossius' ring) concentrically to the curve of the pupillary margin. Vogt noticed in certain cases that the direction of the threads tended to change as they approached the region of the the zonular attachment (equatorial areas). Here they lost their whorl-like aspect and radiated outward in a linear fashion. The nets and starlike figures may often surround an isolated larger pigment clump. Closely packed and condensed, the whole net may resemble moss. Unpigmented grayish delicate maplike designs may result which interrupt the shagreen of the capsule and display a faint color or iridescence. They are usually located peripherally, and maximum pupillary dilatation is essential. Occasionally more intensely white deposits are seen in the form of irregular rounded flat spots with crenated edges, separated from each other by clear areas. Peripheral radiating pigment lines, not unlike the retro-iridal lines described on page 1023, may be found. Larger more ephemeral radial streaks of pigment sparing the axial region are seen in acute or sub-acute iritis after pupillary dilatation. They are composed of fine pigment grains and have been known to become partially absorbed after a month or so. Like retro-iridal lines they may be derived from the pigmented ridges on the posterior surface of the iris, which possibly come in contact with the anterior lens capsule when the iris is swollen (Plate LXXIII, fig. 1).

Another rare finding, described by Elschnig, is the presence of pleatlike formations on the capsule, visible only in the shagreen field. He considered them to be folds of the superficial lamellae of the lens capsule. Since true folds of the capsule can be seen outside of the shagreen fields, Vogt believes the pleats are formed by the separation and folding (?) of delicate extracapsular exudative membranes. (Fig. 431 C.)

Rarely, in association with synechiae, large ropelike or veil-like

PLATE LXXIII

FIG. 1. Radiating pigment lines (resembling retro-iridal lines) and dots following inflammation (iridocyclitis). Early anterior and posterior complicated cortical opacities. Diffuse illumination.

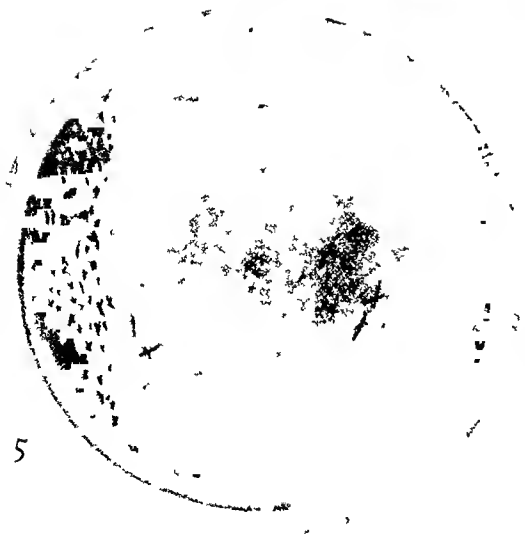
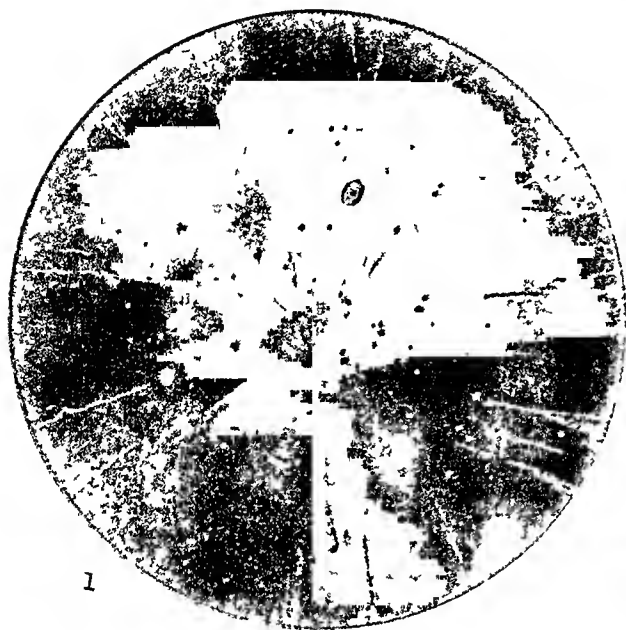
FIG. 2. Same case as shown in Figure 1. Direct focal illumination. Optic section.

FIG. 3. Pigment stars and capsular exudate in iridocyclitis. Note elongated and stretched adhesion extending from capsular exudate to the pupillary border. Diffuse illumination.

FIG. 4. Same case as shown in Figure 3. Direct focal illumination. High power.

FIG. 5. Pigmented stars and exudate on the anterior lens capsule following inflammation.

FIG. 6. Fine netlike exudates and pigment on the anterior lens capsule after iritis.



membranes may occur; these tend to run concentric to the pupil. (Plate XLIX, fig. 4.) They may be connected to the lens capsule or to the pupillary edge. In the beginning such membranes may have been attached to the anterior lens capsule and later are partly or completely detached by the action of the pupil so that one end may remain attached to the pupillary edge of the iris while the other may float freely. In other cases one side or edge of the membrane may remain permanently attached to the capsule and the other to the iris margin. With pupillary contraction such membranes tend to fold, while with dilatation they stretch out.

Exudative membranes in the pupil, especially those attached to the lens capsule and adherent to the iris (e.g., after severe recurrent attacks of iritis or chronic iritis), are frequently subject to vascularization. (Plate L, fig. 8.) Since the vessels do not meet severe resistance they arborize and loop freely in a manner not unlike the conjuncival or superficial corneal loops (in contrast to deep vascularization of the corneal stroma, where they are stubby and run in a direction parallel to one another). As in the cornea fine capillaries are best seen by retro-illumination. With this form of illumination their vascular anastomosing and looplike character is unmistakable. In the light reflected from the deeper parts of the lens, they appear faintly yellow. In direct focal illumination, reflexes arise, causing the appearance of an irregular maplike design on the capsule, which could easily be mistaken for surface irregularities in the membrane containing them. This is especially so when viewed in the shagreen fields. Larger vessels extending from the iris (in seclusion of the pupil) over an exudative pupillary membrane appear bright red by all forms of illumination and offer no diagnostic difficulties (Plate XLIX, fig. 7 and Plate L, fig. 8.)

INFLAMMATORY DEPOSITS ON THE POSTERIOR CAPSULE

In inflammatory states not only of the anterior ocular segment but also of the posterior segment as well, it is not uncommon to find precipitates (deposits, exudates and pigment) on the posterior capsule.

Likewise such deposits are generally seen in "old" cases of retinal separation and occasionally in higher grades of myopia. Vogt found pigment deposits on the posterior lens capsule in a case of retinitis pigmentosa. Before the days of biomicroscopy the detection of such small deposits was not possible.

Precipitates on the posterior lens capsule correspond to the keratic precipitates found on the posterior corneal surface. It may be well here to emphasize that ordinarily the type of precipitates found on the posterior corneal surface and on the posterior surface of the lens are not seen on the surface of the iris or on the anterior capsule. However it has been pointed out that precipitates in the pupillary region of the anterior lens capsule may occur in sympathetic ophthalmia (Koeppel, Vogt and Meesmann). It is claimed that in inflammation, deposits on the posterior capsule are not as frequent as those on the posterior corneal surface. This assumption, I believe, is due to the fact that during the acute phases, owing to corneal and anterior chamber haziness, this part of the lens is not so easily seen. Later, because of their ephemeral character they are sparser than those we are accustomed to find on the posterior corneal surface, but at the same time it is rare not to find some evidence of them. Another factor is that even in the beginning of an attack of iritis or cyclitis or during the quiet phases precipitates may be overlooked easily unless the posterior capsule is sharply focused with the narrow beam and higher powers of magnification employed. This is not always feasible. Needless to say, extensive opacification of the lens, e.g., cataracta complicata or nuclear cataract, may prevent or interfere with accurate observation of minute deposits on the posterior lens capsule. But in spite of such obstacles it is surprising at times how often it may be possible to visualize the posterior capsule with the sharply focused beam. Despite the convex direction of curvature of the posterior lenticular surface, deposits are found predominantly in the lower parts of the capsule. The role played by convection currents in the anterior chamber in the distribution of keratic precipitates would of course have no bearing on the deposition of exu-

dates on the posterior capsule. Here, one could only surmise that their location could be governed by eye movements and gravity. This does not mean that deposits are only found in the middle or

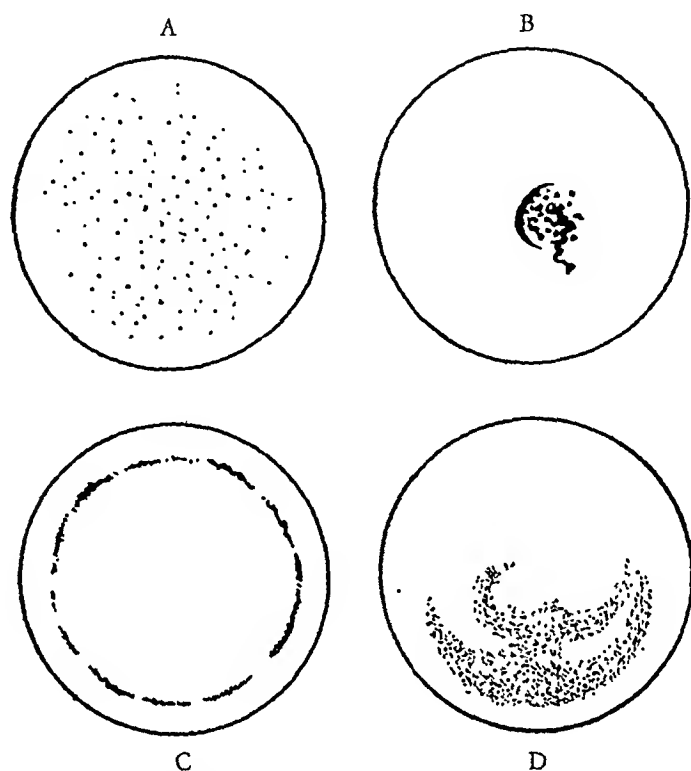


FIG. 433. Types of deposits on the posterior lens capsule. A. Disseminated. B. Perihyaloid. C. Annular. D. Inferior. (After Kobay.)

lower parts of the capsule, for occasionally they may be seen in the upper regions as well (Fig. 415).

With the method of exact localization by means of optic section, deposits on the posterior lens capsule, although commonly associated with vitreous opacities, can be differentiated from them by the fact that the former are stationary, while the latter move with oscillations of the vitreous. Deposits on the posterior lens capsule may appear as small angular dots, delicate lines, larger flat disks, or as a fine whitish dust (Fig. 433). Pigment is also seen as dustlike deposit either disseminated or in circular groups, or as clumps or bands stretching over a considerable area. The pigment varies in color from yellow to reddish brown. Similar to the vitreous in late stages, it may not always be possible to differentiate uveal pigment from

blood pigment (page 1450). As in the case of deposits in the anterior chamber and vitreous, general rules cannot be laid down concerning the types of inflammation in which pigment deposits will predominate over white exudates or vice versa. They may of course be found coexistent, one or the other predominating. Probably, pigment dispersal is more likely to occur when the involvement is of such a nature as to cause swelling and breakdown of the cells containing it.* Pigment on the posterior capsule seems to occur with greater regularity when the iris is markedly involved and after retinal separation. It probably depends on the acuteness, intensity, and localization of the changes. Generally pigment deposits will be found after acute attacks of glaucoma, especially in those cases in which there has been surgical intervention, and in consequence the posterior capsule of the lens can be inspected more easily via the coloboma. In the so-called "quiet" cases of tuberculous iridocyclitis, it is not unusual to find whitish exudations on the posterior lens capsule and in the vitreous. Such small whitish spots, dust, or lines are not always ephemeral in character and rarely may be found even in the absence of corneal precipitates at the outset of the process as well as later. The importance of careful examination of the posterior lens capsule and anterior vitreous biomicroscopically in these cases is self-evident. In cases in which the deposit is large and disklike it is not rare to find bands of vitreous attached to it. Vogt described a case in which there was a large massive exudate with fine radiating white linear extensions in the region of the lental insertion of the hyaloid artery. The whitish exudate had vessels extending posteriorly to the disk. Nasally there were large pigmented belts on the posterior lens capsule extending from the periphery to the white exudate. An area of complicated cataract was adjacent to the white mass. Vogt attributed this to fetal inflammation in the region of the insertion of the hyaloid artery to the lens.

Koby⁵⁰² has pointed out that the topography of the deposit on the posterior lens capsule is determined by Cloquet's canal, the liga-

* Recently I saw a case of Retinitis Pigmentosa in which a complicated cataract was present at the posterior pole. There were numerous pigment deposits on the posterior lens capsule (see Plate LXXXI, fig. 5).

ment hyaloideo-capsulare of Wieger,* and at times by the insertions of the zonular fibers (zonular lamellae). Accordingly, several types of distribution deposits on the posterior lens capsule can be differentiated: (1) disseminated or scattered form; (2) striaform; (3) grouped: (a) perihyaloid, i.e., about the hyaloid insertion, (b) inferior — localized in the lower regions, (c) circular — usually in the form of a peripheral arc (Fig. 433).

Disseminated Type. Depending on the severity of the inflammation the size of the deposits and their distribution varies. In some cases, the deposits may be spread uniformly over the posterior lens capsule as far as accessible to biomicroscopic examination. In others, the distribution may not be so regular, and the accumulations may be greater in the lower portions of the posterior lens surface. The character of the deposits varies chronologically. At the outset, small whitish dots may be discerned, later on in the course of the disease, transformation into stellate figures with processes may be seen. Sometimes by elongation of the processes a spider-web formation occurs. Both these forms may coexist. The tendency toward pigmentation is much less than in keratic precipitation. As in the case of keratic precipitates no inferences can be made concerning the etiology. However, the deposits in chorioretinitis are inclined to be smaller and more often pigmented than those seen in anterior uveitis.

The *striaform* deposits on the posterior lens surface (Koby) have been principally seen following trauma — either contusions or perforation injuries. These striae are suggestive of those seen in the anterior chamber following injury (Vol. I, page 572). In both instances (i.e., anterior and posterior) fibrinous whitish striae may follow upon the posttraumatic hypotony. Such exudation may occur even in the absence of hemorrhage. They are characterized chiefly by their ephemeral nature, those in the anterior chamber clearing up within a matter of hours, while those on the posterior lens capsule resolve more slowly. Not having a double reflecting surface, they can be easily distinguished from folds of the posterior

* It should be noted that this ligament is not demonstrable biomicroscopically. However, anatomically a weak line of adherence of the vitreous to the posterior lens capsule has been demonstrated as a circular zone 8 to 9 mm. in diameter about 1 mm. from the periphery.

lens capsule. As a rule the striae on the posterior lens capsule are much smaller dimensionally than those in the anterior chamber. Koby describes their mossy appearance on the posterior lens capsule.

In the *peribyaloid type*, clumping of deposits occurs within the concavity of the arcuate line around the insertion of the vestigial attachment of the hyaloid artery. Both Vogt and Koby have reported instances of yellowish granular or crystalline deposits in this area. These were associated with macular changes. In the first of Koby's two cases the deposition followed a massive vitreous hemorrhage.

Such deposits may have to be differentiated from congenital or complicated lenticular opacities localized in this area. The optic section reveals the epicapsular location of the former as contrasted with the intralenticular position of the latter.

Inferior Type. One is more liable to find this type following injury where hemorrhage has been sufficiently massive to allow the blood to gravitate inferiorly. In such cases one assumes that the erythrocytes infiltrated behind the posterior lens capsule within the hyaloid vitreous (Berger's space), and were confined below by the annular attachments of the ligamentum hyaloidea-capsulare. Although inflammatory deposits can gravitate to the lower part, massive accumulations comparable to those occurring after hemorrhage have not been detected biomicroscopically.

Circular Form. Pigment deposits can occur in a circular form in inflammatory states either as a continuous or a broken line. Such circular deposits are usually located between the middle and external thirds of the capsular radius. Our chief interest with this form is mainly in high myopes where it may be associated with Krukenberg's corneal spindle. It is still a moot point whether such formations are congenital, postinflammatory, or degenerative. At any rate, cases are seen occasionally in which coexistence of a spindle-shaped pigmentation on the posterior corneal surface and an arc or ring of pigment deposited on the posterior capsule occurs. Because of the finding of pigmentation on the zonular fibers, Koby held that in these cases the annular shape of the pigment deposit is less influenced

by the ligamentum hyaloidea-capsulare than by the posterior part of the zonular apparatus (lamella and fibers). Hence the pigment would be contained in Hannover's canal.

The pigment granules are yellowish or brown and very fine. Concentric annularly scattered grains may also be seen over other portions of the capsule and in the perihyaloid area as well.

EXFOLIATION OF THE LENS CAPSULE

Exfoliation of the superficial layers of the anterior capsule may occur in four conditions: * (1) traumatic; (2) toxic (in atrophic eyes, after prolonged iridocyclitis or from the effects of a retained foreign body in the lens); (3) action of heat (infrared) (page 1295); and (4) senile (with or without glaucoma capsulare or cataract).

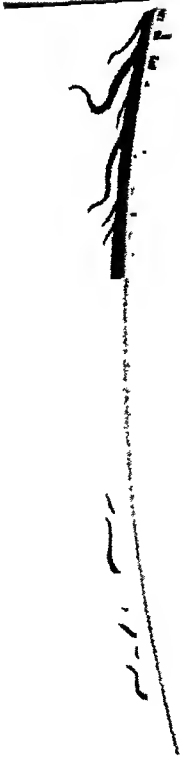
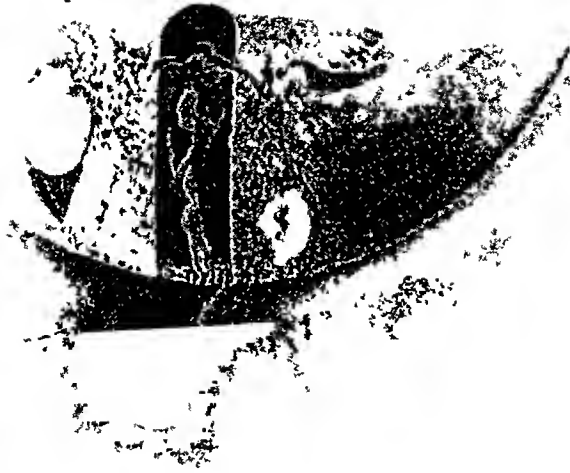
In this chapter only the so-called "senile" exfoliation of the anterior lens capsule will be considered. According to Busacca (1927)³⁷⁸ the lens capsule consists of three main divisions: (1) The overlying zonular lamella, derived from the terminal expansions of the zonular fibers. This occupies the equatorial portions of the capsule in an area 2 mm. wide thus comprising approximately one-quarter of the total capsular surface. (2) A pericapsular membrane which covers the entire lens; and (3) the main part of the capsule which, as has already been pointed out, may be composed of several layers. Since in exfoliation of the lens capsule and in heat cataract the separation may occur in the pupillary part as well as peripherally, it follows that at least in the case of its more central location that the zonular lamella is not involved. Actual separation of the zonular lamella has been noted in other conditions with the biomicroscope by several authors (page 1208). In these cases a membrane detached from the equator was seen having zonular fibers inserted into it. In heat cataract the separated lamellae are many times thicker than in senile exfoliation. Consequently in the former the detached membrane is clear and lustrous and does not show the granular fragmentation and destruction so characteristic of senile exfoliation. In heat cataract, the thicker separated membrane ("fire" lamella) hangs as a

* Modified after Duke-Elder.

continuous glasslike structure (comparable to that seen in separation of Descemet's membrane [Vol. I, fig. 203]), its sharp free edge tending to roll up in the anterior chamber axially. This free or floating curled edge (in the anterior chamber) is directed toward the periphery, whereas in senile exfoliation the detached edge is turned axialward. In heat cataract the axial part of the lamella is generally adherent to the underlying and remaining hyaline capsule, causing an opacification. The separation from the capsule, the site of which is probably near the equator, is gradual and is not marked by any sudden visible change. Another distinguishing feature of heat or fire cataract is the absence of the secondary glaucoma, frequently seen in senile exfoliation (glaucoma capsulare).

The fact that a peeling or exfoliation of the superficial layers of the hyaline lens capsule does occur clinically confirms the anatomic finding in maceration preparations that this structure is laminated. This is especially noteworthy in those cases in which it has been seen biomicroscopically that in one place successive layers may become exfoliated. That is, after the most superficial layer is cast off, the layer beneath may likewise start to peel. Exfoliation of the superficial layers of the lens capsule (senile type) rarely occurs before the sixtieth year (however, a case occurring in a 41-year-old woman was reported by Gradle and Sugar ⁴⁵⁷). In 1917, Lindberg ⁵²² mentioned the finding of white flakes deposited on the pupillary border in glaucoma. In 1918 Vogt ⁶¹⁸ described the case of a 72-year-old man who later developed glaucoma. He mentioned the presence of a film on the anterior lens capsule, the edges of which were crinkled. Because of the presence of pupillary threads Vogt considered that the change was associated with residua of the pupillary membrane. Two years later he saw another case with chronic glaucoma, in which the film appeared as a faint limited capsular disk with crenated edges within the pupillary area. There were numerous bluish-white feltlike or crumblike deposits at the pupillary margin of the iris. Because this was found postoperatively he thought that they might be inflammatory products and stated that perhaps both structures, i.e., on the lens capsule and iris, consisted of the same substance. In

A



B

FIG. 434. A. Exfoliation of the superficial lamella of the lens capsule. Note particles on the pupillary margin. B. As seen by optic section.

PLATE LXXIV

FIG. 1. Senile exfoliation of the superficial lamella of the anterior lens capsule. Direct focal illumination. Note senile atrophy of the iris as viewed by retro-illumination. Exfoliated particles on the iris surface.

FIG. 2. Senile exfoliation of the superficial lamella of the anterior lens capsule. Case of glaucoma capsulare. Illustrating exfoliating dandruff-like particles some of which are attached to the pupillary margin of the iris.

FIG. 3. Senile exfoliation of the superficial lamella of the anterior lens capsule. High-power view. Edges of exfoliated layers are curled back before breaking off.

FIG. 4. High power view showing details of exfoliating areas. Surface beneath exfoliating layer is beginning to desquamate.



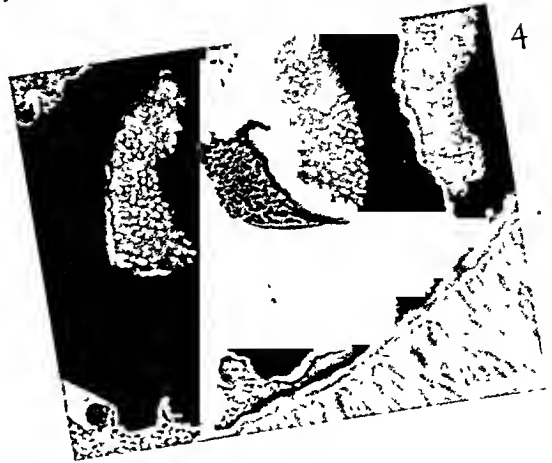
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3



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4

1923, he came nearer the truth when, after finding the condition in cases of glaucoma that had not been operated on and were quiet, he postulated that both the capsular change (grayish disk with crinkled edges) and the deposits on the iris were not of inflammatory origin but resulted from an exfoliation of the superficial capsular lamella. This conclusion was strengthened not only by the physical appearance of the film (crinkling and eversion of its edges) but also because of the fact that exudative membranes do not tear off or crumble, no matter how thin they are; nor would they be only confined to the anterior capsule and be distributed in such a uniform way.

The exfoliated particles at the seam, or the iris surface or more rarely on the posterior corneal surface were never pigmented. There were no signs of iritis and the flakelike deposits themselves had none of the characteristics of inflammatory exudate. Hence, it was concluded that these grayish feltlike structures, especially characteristic for the pupillary seam or margin, are of the same nature as the material seen on the lens capsule but were deposited secondarily (Fig. 434). It may be that the pigmented excrescences with their uneven and granular surfaces favor the adherence of the particles swept off from the anterior lens capsule by pupillary action.

The central disk in the pupillary area may be very faint and may be easily overlooked. However, in most cases its periphery is outlined by a more opaque ring or edge of exfoliated material. Frequently the ring is seen, but the delicate film within it is missed unless the pupil is dilated. After this is done, one can compare the grayish color of the central disk with the adjoining area which, being film-free, is darker.

Dilatation of the pupil to a width* of at least 4 mm. shows in nearly all cases a second phenomenon, i.e., the presence of a more opaque peripheral wreath of similar but more marked granular-appearing structures or projections, arranged in a radiating fashion, located in an intermediary zone between the periphery and the central disk (See Plate LXXIV, figs. 1, 2, 3, 4). The area between the disk and the more peripheral grayish deposits are clear, although

* Failure to dilate the pupil and to employ the biomicroscope is probably the reason why many cases of capsule exfoliation are missed. This is understandable since from 50 per cent to 75 per cent of cases having this condition are associated with glaucoma.

occasionally a few flakes may be deposited on it. The filmlike nature of the grayish material is best demonstrated at the edges, where exfoliation causes crinkling or even eversion. According to Vogt one gets the impression that there is a delicate membrane formed over the entire anterior surface of the lens and that at certain sites, i.e., where the posterior surface of the iris is in contact with the lens normally, this membrane seems to disappear either partially or completely. It should be recalled that the relief of the posterior pigmented iris surface is formed of radiating ridges (page 734). Where these ridges contact the lens capsule, they could either prevent the deposition on the capsule, or owing to sweeping movements, could keep this area clear of the exfoliated product. Iris movements, like a broom, could sweep away the granular degenerated products of the separated lamellae and leave clear areas behind. The absence of the possibility of such friction on the posterior capsule (if the condition does occur there) may account for the fact that granular accumulation on this part of the capsule has not been seen.

In contrast to the granular aspect of the film peripherally, the surface of the central pupillary disk is generally smooth. Except for the fact that its center (pupillary area) never contacts the iris, it is difficult to explain why this area does not become as granular as those in the periphery. However, the edges of the smooth central disk may show crumblike granules in which case a ring is formed. This may result from action of the pupillary margin which may "sweep" up the particle in this region.

Another point to be considered in the formation of exfoliation is the action of the aqueous. Once a rupture has occurred in the superficial lamellae entrance of this fluid might cause further separation and degeneration. Repeated observation of an individual case will reveal not only changes in the arrangement but also even the disappearance (perhaps to the angle of the anterior chamber) of the exfoliated flakes, not only those of the lens capsule but also those on the iris seam. As mentioned before, successive layers of the capsule can come away, so that new exfoliated particles may appear as time

goes on. Following iridectomy the peripheral parts of the anterior capsule can be inspected.

It will be seen that the exfoliative changes decrease toward the equator. The radial extensions of the film become granular up to the region of the zonular insertion, where it (the film) tends to separate in a concentric manner. In the region of the zonular insertion, delicate whitish parallel lines (zonular fibers) can frequently be seen. As Vogt has shown, the detachment of the lamellae at this point is not fortuitous but could result from the traction of the zonule, the force of which would tend to tear the film or pellicle at right angles to its direction, or concentrically.

Another rather interesting finding in this condition was that of Vogt who on several occasions noted the sudden appearance of a cloud of pigment in the anterior chamber upon dilatation of the pupil with cocaine. None of these patients had diabetes but all had chronic simple glaucoma. The question arises whether this is related to separation of the superficial lamellae or whether it resulted from pigment changes known to occur in prolonged pilocarpine miosis (page 787). However, he noticed it once in a case of glaucoma which was not complicated by senile exfoliation of the superficial lamellae and also once in a case (senile exfoliation and glaucoma) in which no miotics had been employed. In addition to resulting from a sudden bursting of iris pigment-containing cells, it might also occur from an accumulation of stored pigment in the posterior chamber. Dilatation of the pupil would permit the pigment to pass into the anterior chamber. Vogt found that following such dilatation a small cloud of pigment appears, sometimes tongue-shaped, in the anterior chamber in the region of the upper pupillary margin. In a few minutes it dispersed in the anterior chamber. In one case after half an hour there was only some fine dust to be seen. On further dilatation another cloud appeared and he found four small radial stripes of pigment on the anterior capsule. The second cloud was dispersed in a few minutes. The constant finding of pigment (and exfoliated particles) in the chamber angle gonioscopically (Barkan, Gradle and Sugar) in cases of exfoliation of the capsular lamellae might indicate that the sud-

den presence of pigment in the anterior chamber as described above is not surprising and that this phenomenon is probably considered rare only because few have observed it biomicroscopically. Considering the advanced age of most of the patients pigmentary degeneration of the iris is understandable. Gradle and Sugar (examining Terry's material) found serrations on the pigmented posterior surface of the iris characteristic of capsular exfoliation. According to Sugar (Vol. I, chap. 17) pigment granules deposited in the trabecular spaces are derived from the pigment epithelium of the iris which is traumatized by its constant movements against the roughened superficial capsular lamella. Irvine confirmed the finding of Trantas, Hörven, Gradle and Sugar that exfoliative material is present on the zonule in iridectomized eyes. This raised the question whether the deposits were secondary, i.e., from the lens capsule, or whether a degeneration of the zonule *per se* also occurs (Irvine, and Gradle and Sugar). Basing their contention on the possibility of a primary zonular disease, these writers also emphasized the commonly found vitreous changes (degeneration and fluidity), and suggested the idea that in exfoliation of the capsular lamellae we may be dealing with a syndrome involving many parts of the eye.

Considering that on the capsule the exfoliated material, principally, is not derived from the zonular lamella but from the superficial lamellae of the capsule axially to the insertion of the zonule, it would seem that the former supposition (i.e., that the deposits on the zonule, chamber angle, iris, and rarely the cornea are secondarily derived from the exfoliated material of the capsule) is true. Taking into consideration the age of the patients having this condition, I recently reexamined a case of capsular exfoliation in which there were no signs of glaucoma and compared her vitreous with that of another presumably normal elderly person of approximately the same age. Both showed the typical changes of the aged vitreous to about the same degree.

Glaucoma Capsulare. This term was coined by Vogt to designate the secondary glaucoma that so commonly occurs as a complication to exfoliation of the capsular lamella. (See Vol. I, page 628). The

frequency of this association (probably close to 75 per cent) is demonstrated by Irvine's tables (see Tables XIX and XX).⁴⁸⁵ Originally both Lindberg (1917)⁵²² and Vogt⁶⁵⁹ thought that the exfolia-

TABLE XIX
PERCENTAGE OF PATIENTS WITH EXFOLIATION
HAVING GLAUCOMA (AFTER IRVINE)

AUTHOR	YEAR	NUMBER WITH EXFOLIATION	PERCENTAGE WITH GLAUCOMA
Vogt ⁶⁶²	1931	45	75
Busacca ³⁷⁶	1927	30	90
Trantas ⁶³⁹	1929	42	33
Rehsteiner ⁵⁷⁴	1929	78 (from the litera- ture)	64
Grzedzielski ⁴⁶²	1931	156 (from the liter- ature)	58
Baumgart ³⁵⁷	1933	46	63
Sobhy ⁶¹²	1932	24	54
Alling ³⁴⁶	1927	7	14
Kirby ⁴⁰³	1930	7	29
Holloway and Cowan ⁴⁷⁹	1931	3	67
Garrow ⁴⁴⁵	1938	10	80
Irvine ⁴⁸⁴	India	18	44.
Irvine ⁴⁸⁴	Los Angeles	8	50

tion was a phenomenon secondary to simple chronic glaucoma. Later Vogt found many cases of exfoliation without glaucoma, and cases in which glaucoma developed during the period of observation, and he was forced to the conclusion that the glaucoma was secondary. According to Vogt's supposition (later substantiated by gonioscopic studies) particles of exfoliated material after a time become heaped up in the filtration angle, and eventually block it. He drew attention to the observation of Hess that large amounts of exfoliated capsular material (cuticular) would be material difficult to dissolve, digest,

or absorb. The presence of exfoliated particles and pigment on the trabeculae was later confirmed gonioscopically by Barkan,³⁵³ and Gradle and Sugar⁴⁵⁷ and histologically by Sobhy Bey (1932).⁶¹²

TABLE XX

PERCENTAGE OF PATIENTS WITH CHRONIC SIMPLE GLAUCOMA HAVING EXFOLIATION (AFTER IRVINE)

AUTHOR	DATE	NUMBER WITH GLAUCOMA	PERCENTAGE WITH EXFOLIATION	COMMENT
Lindberg ⁵²²	1917	60	50	Pupils not dilated
Vogt ⁶⁶²	1931	150	9	
Malling ⁵⁸¹	1923	81	40	
Baumgart ³⁵⁷	1933	59	49	Pupils dilated
Hörven ⁴⁸²	1937	150 (not operated on)	85	Coloboma present
.		43 (not operated on)	93	Pupils dilated
Garrow ⁴⁴⁵	1938	51	16	Pupils not systematically dilated
Irvine ⁴⁸⁴	India	33	24	Pupils dilated
Irvine ⁴⁸⁴	Los Angeles	125	4	Pupils dilated in only 26 cases; percentage therefore low

Sugar also observed that in glaucoma capsulare peripheral synechiae are relatively absent. Hörven,⁴⁸² advanced the idea of “an altered permeability” of the zonulocapsular diaphragm as the factor in the causation of glaucoma. Irvine’s observations⁴⁸⁵ tend to confirm this view.

Chapter Twenty-Seven

INJURIES TO THE LENS

THIS chapter includes discussion of traumatic lesions resulting from contusions and perforations, and the lens changes produced secondarily by retained intra-ocular foreign bodies (especially iron and copper). Since biomicroscopically the picture of traumatic luxation and subluxation does not differ essentially from that seen in spontaneous (hereditary) displacements, both depending on pathologic changes in the zonule, they are considered together in the chapter on the zonule (page 1345) and on page 1206 under the heading of complicated cataract. In a great many instances the morphologic aspect of the changes provoked by the above-mentioned causes are so characteristic that it is possible today in many cases with the aid of the biomicroscope to differentiate them from other types of cataract. The value of accurately recorded biomicroscopic examinations in traumatic cases hardly needs to be emphasized here. One cannot, however, rely on the morphologic appearance of lens changes alone. In addition to a thorough survey both of the functional and ophthalmoscopic aspects, the biomicroscopist carefully examines the conjunctiva and sclera (for congestion, hemorrhages, scars, etc.), the cornea, the anterior chamber (depth and flare), pupil and iris and vitreous because it is rare that trauma sufficient to produce lens changes does not cause other changes, especially in the anterior segment. This holds not only for fresh cases, in which the history of an injury is incontrovertible, but it applies also to cases involved in medicolegal disputes years after a reputed injury. Again the importance of the use of the narrow beam for exact localization of lens changes should be emphasized. With the unfocused beam or even with the focused wide beam a pan-

oramic view of an opacity may be obtained, but exact localization is almost impossible. This is particularly important when it is necessary to know whether a change is located in the capsule or just below it. When employing the focused wide beam, angulated so that the mirror zone (specular reflection) lies partly over a subcapsular opacity, it may be possible to determine that the shagreen of the unaffected anterior capsule lies in a plane anterior to the lesion. But with the narrow beam the illuminated capsule and anterior band of disjunction (the latter cannot be seen with the broad beam) are seen as sharp lines and exact localization becomes relatively simple.

In most of the discussions on cataract due to trauma, the subject is considered from the standpoint of the presence or absence of capsular rupture. Cases in which even the biomicroscope fails to reveal any rupture of the capsule are called "contusion" cataract. They may follow direct or indirect blows to the eye, with or without evidence of rupture or perforation of the outer ocular tunic. The possibility of submicroscopic openings in the capsule in contusion cataracts has not as yet been disproved. Ruptures in the neighborhood of the equator in the absence of a coloboma, or even of the posterior capsule in the presence of extensive cataractous change, could elude biomicroscopic detection. The pathogenesis of traumatic cataract following frank rupture of the capsule (e.g., by the passage of a foreign body or by the penetration of sharp objects) in which the aqueous can gain access to the lens fibers through a gaping wound, is not difficult to understand. However, in contusion cataract where no apparent capsular perforation has occurred, the mechanism involved in its production is as yet not clear. Several interesting hypotheses have been advanced to explain the mechanical basis of the lens opacities seen in contusions.

Vogt drew a parallel between contusions of the eyeball and the skull (about which more is known). The outer wall of the eyeball is of course softer and more elastic than the bony skull. (The softer skull of the newborn and of children would perhaps offer a better comparison.) Hence when force is applied (but not of sufficient strength to cause an external rupture, or fracture), its motion will be transferred to the interior and will cause a shifting or "a gliding" of the contents. As a result of the acute increase of internal tension, the pressure effects (in the

case of the brain [Kocher]) may set up a condition corresponding to commotio and at the same time may provoke a strong contusion in the direction of the blow. In the eye such effects as Vossius' ring or Berlin's edema may result. According to hydrodynamic laws, the contents of the skull and eyeball being considered as fluid, the effect of the force is transferred not only in the direction of the blow, but with equal intensity in all directions. Therefore, internal pressure develops which acts from the center peripherally so that the contents may be hurled against the wall; depending on the intensity of the external force, varying degrees of damage may result. In the eye this might result in tears of the sphincter, iridodialysis, capsular tears, luxation of the lens, tears of the retina and choroid. According to Vogt, in *contusio bulbi* the most important role is played by the "throwing motion" of specifically heavy parts. The specifically heavier iris will be pressed for a moment against the lens, thus producing Vossius' pigment ring on the capsule. The lens can be broken loose from its zonular attachments and forced posteriorly into the vitreous. If the eyeball is ruptured, the lens may be extruded. In the posterior segment of the eye, the pressure of the vitreous may bore a hole in the macula and when the retina is thrown against the sclera, Berlin's opacity can result. Although a considerable amount of the force is dissipated when an actual perforation occurs, still a wave of high pressure may also set up further damage in parts not in the direct line of the force. An example of this was seen in a case of a perforating injury when a Vossius' ring formed.

Frenkel, who considered the effects of contusion in the anterior segment as a syndrome, believes that the motion imparted to the lens as a result of the force is the primary factor in causing the different alterations seen in the syndrome. Under the impact, the lens is pushed back against the vitreous (the zonule often being partially torn). On the rebound it snaps forward and "distends the iris diaphragm like the head of a fetus against the neck of the uterus." As a result (of the force?) of the recoil, damage to the lens and iris occurs. The following are the lesions of the anterior segment syndrome in the order of their severity: (1) paralytic mydriasis (isolated, often transient); (2) internal ophthalmoplegia; (3) lesions of the iris—small pupillary ruptures, minimal iridodialysis (often not easily seen); (4) localized subcapsular opacities; (5) subluxation of the lens. In addition Frenkel calls attention to the following details which must be looked for carefully with the biomicroscope: (1) careful examination of the depth of the anterior chamber; differences of level is often the only sign of subluxation of the lens; (2) noninflammatory rests of iris pigment on the anterior surface of the lens (including Vossius' ring); (3) localized lens opacities (which may be due to tearing of the capsule? and penetration of aqueous [these opacities may be stationary or, rarely, may become progressive]); (4) a semicircular concentric fold of the iris. This sign is of importance in nonapparent subluxations of the lens.

Frenkel also differentiates between traumatic syndrome of the anterior segment in the case of direct action on the globe and traumatic syndrome of the posterior segment in the case of indirect action. It should be pointed out that the symptoms of anterior segment contusion are more conspicuous in adults than in the young.

This has been explained by the suggestion that in the young, owing to the greater elasticity of the iris and molding power of the lens, the brunt of the contusion is more likely to be borne at the retinal-zonule insertion than at the lenticular-zonule insertion.

Other authors have suggested that the cause of contusion cataract lies in fact that owing to either alterations in the capsular epithelium directly or variations in osmotic pressure of the aqueous, fluid gains entrance into the lens. The fact that rosette-shaped opacities are so common leads to the opinion that the suture system of the cortex is the locus minoris resistentiae (Vogt).

CONTUSION CATARACT

That certain characteristic types of lens opacities are peculiar to nonperforating contusions (some of which were described before the days of biomicroscopy) is now generally agreed. Biomicroscopic examination, however, has been of immeasurable value in confirming previous impressions, because by its means not only can the lesions be seen better, but what is of greater importance, they can be accurately localized and differentiated from opacities due to other causes.* In addition it is possible now to follow accurately the progress of the lens changes and to note minute details of their morphology. It is known that in certain cases delicate opacities (probably due to edema) appear immediately after a contusion and may later disappear—in other words, they represent reversible phenomena. However, in most instances the opacities resulting from contusions are permanent but nonprogressive, although in the case of the subcapsular alterations, their localization may change. Localized opacities in the subcapsular region (the most vulnerable part of the lens—see *cataracta complicata*) can be displaced deeper into the cortex by the ingrowth of new fibers (Vogt).

As already mentioned, the biomicroscope also assists in detecting

* Davidson³⁹⁴ states: "The importance of the use of the narrow beam and of retroillumination cannot be too much stressed. It has served to rectify some previously held notions. It shows a much greater rarity of posterior-capsule ruptures, an intactness of the equatorial capsule when accessible to examination in colobomata, and draws our attention to the biochemical factor alongside the mechanical factor of microscopic solution of continuity, in the production of a contusion lens opacity. It has also established the difference between contusions in the young and in the adult in their effect on the lens. It has demonstrated that an opacity has the tendency to traverse the equator backwards along the same discontinuity surface or zone, and that zonular, coronary opacities and riders and other cortical opacities can be observed in contusions."

confirmatory signs of trauma especially in the iris and vitreous, the importance of which has been pointed out by Frenkel. Besides the more characteristic traumatic lens opacities, e.g., Vossius' annular (ring) opacity and the traumatic rosette opacity, forms identical to typical presenile and senile lens changes (cortical punctate, lamellar and zonular, coronary, cerulean, nuclear and posterior saucer opacities) have been described and attributed to the effects of contusion. Such an assumption must be valid in the face of incontrovertible history of injury, signs of anterior contusion syndrome (more common in older persons) and provided the lens in the uninjured eye was unaffected — since ordinarily, when of developmental origin, these forms tend to be bilateral. Davidson has also pointed out that injury may activate the progression of pre-existing presenile and senile cataractous changes. As with complicated cataract from other causes the morphologic appearance of contusion cataract may depend on the age of the individual. So that in the aged, whatever the cause, the resulting lens opacity may resemble that of developmental senile cataract. This point, against which there seems to be no *a priori* objections, requires further confirmation; if further verified, it would be of great importance in medicolegal cases. From an analysis of 57 eyes with contusion lens opacities (in persons of varying age) Davidson indicates that the typical or characteristic types of contusion opacities, i.e., capsular (Vossius' ring opacity), subcapsular (nondistinctive or rosette forms), equatorial (lamellar) and posterior-lens involvement-tendency, and regional segment-like localization are morphologically specific for the young. After 45 years of age senile pathologic changes intervene and hence the specific morphology of contusion cataracts after this age becomes less discernible. That such variations should occur is conceivable when one considers the variances in the morphology and development of the lens during life. But as Davidson states: "Any generalizations in regard to specific morphologic picture of contusions of the lens at different age periods must, in the nature of things, be regarded as provisional." In the end one must always consider that the nature and force of the blow varies in every contusion and that as

PLATE LXXV

FIG. 1. Vossius' ring. Diffuse illumination.

FIG. 2. Contusion cataract. Partial rosette seen twelve years after an injury. Diffuse illumination.

FIG. 3. Same case as shown in Figure 2. Showing deep location of rosette. Late Remotio. Direct focal illumination. Optic section.

FIG. 4. Rosette opacity after perforating injury near the limbus.

FIG. 5. Optic section, same case as shown in Figure 4. Direct focal illumination. Optic section.

FIG. 6. Lamella type of opacity following contusion. Note subcapsular location of the opacities.



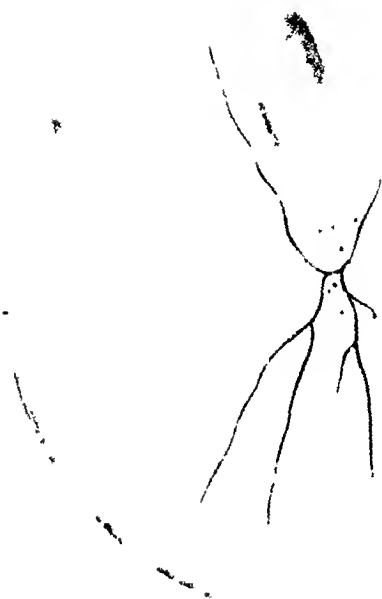
2



5



3



4

6



yet no definite relationship between the degree of force and its results have been established.

The lens opacities, characteristic of contusion, are listed below (it should be remembered that many of them may occur in cases in which the capsule has obviously been ruptured, e.g., perforating injuries) :

1. Vossius' annular (ring) opacity
2. Disseminated subcapsular opacities (Vogt) and their later displacement. Also "button-shaped" opacities (cataracta nodiformis)
3. Traumatic rosette-shaped opacity (recent and late [remotio cataractae traumaticae])
4. Lamellar opacity (zonular)
5. Presenile (coronaria and cerulea) opacity
6. Progression of previously existing senile opacity
7. Posttraumatic atrophy of the lens (Vogt)

VOSSIUS' ANNULAR OPACITY

In 1903 Vossius⁶⁶⁶ described the finding of a ring-shaped pigmented opacity on the anterior surface of the lens following trauma to the eye. In outline the ring runs concentrically to the pupillary margin and generally can be seen in the red fundal reflex ophthalmoscopically. According to Vossius the mechanism by which the ring is formed corresponds to the imprint by a rubber stamp ("Stempelabdruck"). In other words, owing to the effect of the transmitted force, the posterior pigmented surface of a portion of the iris is pressed against the lens and leaves an imprint (Plate LXXV, fig. 1). This means that the ring is formed at the instant of the contusion and that it is not formed by a later deposition, e.g., from pigment in the aqueous. Many of its morphologic and physical characters substantiate this opinion. The ring has been seen almost immediately after contusion. With lapse of time, instead of increasing in size or prominence, it tends to absorb. This has been noticed even when a hyphema has been present for long periods of time. It is true that pigment may be deposited on the anterior capsule in ring

form even in latent iridocyclitis, but in this instance there are morphologic differences: The width of the ring is narrower; the pigment coarser (clumps) and isolated particles are seen peripheral to the ring upon dilatation of the pupil; and usually remains of whitish exudates are found on the capsule to which pigment is frequently adherent.

Biomicroscopically, Hess and, later, Vogt showed that the Vossius ring does not represent an actual opacity of the capsule or lens substance but that it is the result of a thin deposit on the surface of the anterior lens capsule. The ring itself consists of a flat (single) layer of fine pigment granules, usually of a reddish brown or bronze color, often glittering. Depending on the intensity of the light,* the visibility of the ring will vary when observed in the mirror reflex of the anterior capsule. With the lowering of the intensity of the light the shagreen becomes hidden by the ring as the latter passes over it and, conversely, if the intensity of the light is strong with consequent increase in brilliance of the shagreen, the fine granules of the ring may be obscured. The single-layered and flat disposition of the pigment particles (adherent to the capsule) disputes any secondary accumulation, which might be expected if the pigment were deposited or precipitated from the aqueous as in inflammations or hemorrhage. With higher powers of magnification, $40\times$ or more, the concentration of pigment granules is seen to lessen in the direction of the axial and peripheral edges of the ring where their distribution also becomes more irregular. Frequently the ring is not continuous but rather appears to be more or less regularly interrupted or segmented. Here and there small dark gaps or holes (pigment free areas) will be seen. This appearance has been explained by the fact that the posterior surface of the iris (page 734) normally is not smooth but is composed of closely contiguous radial ridges. Law stated that he had never seen a case of Vossius' ring opacity that did not clear completely and permanently.⁵¹⁸

In a case measured by Vogt the over-all horizontal diameter of the ring measured 3.5 mm.; temporally the width of the pigment deposit

* Governed by the width of the beam or by altering the rheostat.

was 1.22 mm. while nasally it was 1.25 mm. Above and below its width varied between these two measurements. According to Vogt it is not clear why the same type of contusion produces a ring in one case and not in another, and why it is seen only in the young and not in the aged. In the latter case he feels that some role must be played by anatomic factors peculiar to the young (softness and elasticity of the tissues). Both Hess and Zentmayer have cited cases to prove that the ring was formed secondarily to hemorrhage in the anterior chamber and its presence is not necessarily conditioned by an antecedent contusion. Vogt stated that he had never observed Vossius' ring formation in cases of nontraumatic anterior chamber hemorrhage. After traumatic injury, blood and pigment may be present in the anterior chamber for a long time, but the ring always appears most intensely on the first day of its observation, and in spite of the presence of blood, the deposits forming the ring tended progressively to disappear. Also, in stressing the fact that the ring develops immediately at the time of injury, he brings out the point that double or multiple ring formation has never been seen after the original one developed. This speaks against a later deposition from pigment or blood in the aqueous. This should be possible if, as postulated by Hess, the deposit consisted of blood pigment. He explained the ring formation as resulting from sweeping together of the blood cells by iris movement. Hence, upon dilatation of the pupil (either secondary to sphincter lacerations or mydriatics) theoretically a second ring could form. In addition, with high power the dark reddish-brown color of the pigment granules of the ring can be noted. One gets the same color impression of the dark iris when examining it under high magnification. Under the same conditions blood cells appear pale yellow.

In explaining the pathogenesis of the ring Vossius stressed the following points: (1) compression of the cornea; (2) desquamation and adherence of the pigment epithelium; (3) degeneration of capsular epithelium and at times also of the underlying lens fibers. Frenkel objected to this viewpoint of Vossius and to that of Wageman and others (i.e., depression of the cornea, displacement of the

aqueous, and distention of the sclero-corneal ring). According to this author the sequence of events in the formation of the ring is: (1) rupture of the zonule; (2) subluxation of the lens; (3) forcing of the lens against the vitreous; (4) rebounding action of the lens against the posterior surface of the iris. Frenkel contends that the zonule is not as easily stretched as is commonly believed but that to the contrary it tears more easily. His idea of the rebounding action of the lens would certainly explain those cases (Steiner, 1910;⁶¹⁸ Purtscher, 1913⁵⁶⁵) in which a ring formed in consequence of intra-orbital extra-ocular bullet lesions causing compression of the globe.

TRAUMATIC DISSEMINATED SUBCAPSULAR OPACITIES (VOGT)

Thin, flattened flakelike or punctate opacities layered or irregularly distributed in the anterior parts of the lenticular cortex may occur as an early or late manifestation of injury (Fig. 435 B). These opacities are usually discrete and tend to be stationary; but later, owing to the growth of new lens fibers, they may be pushed somewhat deeper into the cortex. This is attested by the fact that recent ones, which seem to be attached to or to lie just below the capsule later, become separated from it by a layer of clear lens substance. This fact gives confirmation of the continuous growth of the lens (see Polar Cataract). The genesis of the zones of discontinuity and the development of the intervening layers has always been a baffling problem. In the normal lens with the ordinary nitra light source the uniform opalescence of the layers on both sides of the demarcation line deprives the observer of any means of following the shifting and rearrangement of the different layers which are bound to occur during growth.

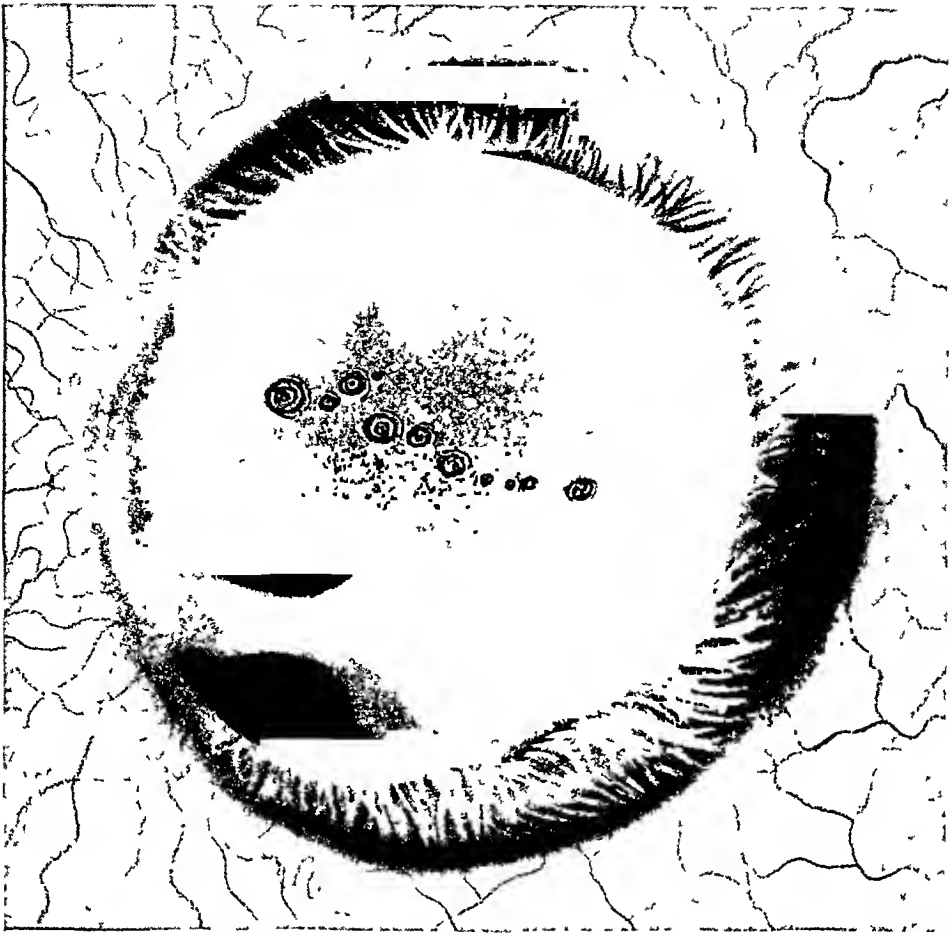
The presence of a localized traumatic cataract offers another approach to the problem of lens growth. These opacities and also the larger rosettes, which are usually thin and sharply defined, can be accurately located with a narrow beam, and since they change not at all or very little in size and shape, any alteration in their location can easily be followed. In the beginning such opacities are situated immediately below the capsular epithelium and later will be

seen pushed deeper and deeper by newly formed lens fibers. The two landmarks for the observation are: (1) the disjunction line; (2) the adult nuclear band. With this method* it is possible to determine (by comparison with the normal fellow eye) if the trauma has been strong enough to interfere with the production of new layers, or to abolish their production entirely. In some instances, as the opacity migrates deeper, tags of it may remain adherent to the capsule, thus offering a confirmation of the interpretation just mentioned.

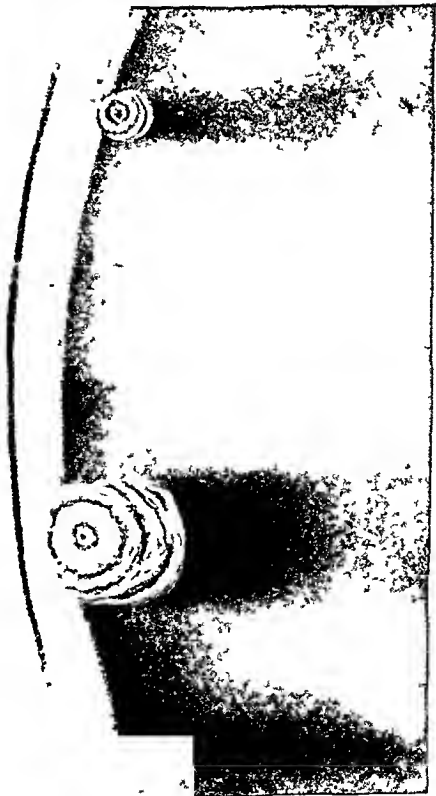
Vogt first observed traumatic disseminated subcapsular opacities in 1922 in the left eye of an 8-year-old boy. This eye had been struck by a chestnut two years previously. At the time of the injury there were ruptures of the sphincter and blood in the anterior chamber. The blood disappeared the following day and a typical Vossius' ring was present. At this time there were no cortical opacities. Two years later, on reexamination, there were numerous white punctate opacities irregularly scattered in more or less the same level with and below the anterior line of disjunction. Vossius' ring was no longer present. The uninjured eye did not show any changes. In this case Vogt considered the opacities to be a late manifestation or secondary complication of the ring. In another case similar opacities were seen in association with a Vossius ring, nine days following a contusion. Three years later the punctate opacities not only persisted but were slightly increased in number. In one case there was a limbal rupture with prolapse of the iris, which was excised. Ten days later in the area of the coloboma numerous small white dots were seen subcapsularly. They did not follow any orderly or radial arrangement, a condition noted by Foster in his report.⁴³⁰ At times coalescence of the flakes may result in a thin flattened continuous layer of opacity with radiating processes (opacities and vacuoles)

* Some resemblance can be found between this method and the method used by geologists to study the movement of glaciers. In the great expanse of ice and snow it is impossible to tell whether there is motion or not. By choosing an immovable landmark like the pinnacle of a mountain and placing an easily recognizable object like a black rock on the glacier, the distance between these two can be measured at the beginning of the observation and through the years, and so, the motion of the glacier can be found and recorded.

A



B



C

FIG 435. A Cataracta nodiformis B. Disseminated subcapsular opacities in the cortex below the anterior line of disjunction. (After Vogt) C Cataracta nodiformis. High power.

having an appearance somewhat like the nontraumatic anterior complicated cataract caused by intra-ocular disease.

A very interesting and hitherto rarely mentioned type of trau-

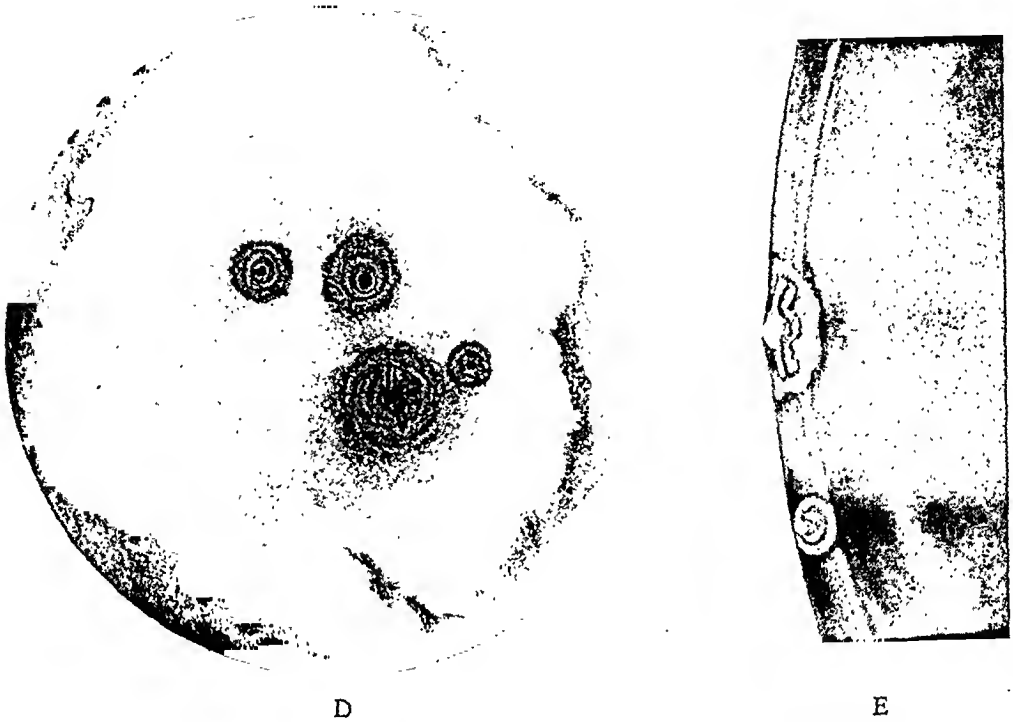


FIG. 435. D. Cataracta nodiformis, diffuse illumination. E. Same case as in D, optic section.

matic subcapsular opacity is the so-called "button-shaped" form (cataracta nodiformis). They appear studlike (collar-button-shaped), rounded, and layered, not unlike small, pyramidal, polar cataracts. Their color is whitish and they tend to develop some time after the injury has occurred. In the case illustrated by Figures 435 A and C the buttons were first noticed two years after a perforating injury. On the day following the injury prolapsed iris tissue was excised. Two weeks later a flattened lamellar opacity was seen in the neighborhood of the coloboma (Fig. 435 A). This did not progress.

When frank signs of previous inflammation (e.g., keratic precipitates or synechiae) are absent, as might be the case in latent forms of iritis or cyclitis, and in the absence of a history of trauma, differential diagnosis may be very difficult. Ordinarily, in compli-

cated cataract the tendency to deeper displacement by clear lens fibers with time does not occur. Whereas complicated cataract tends to progress, localized contusion opacities, in contrast, are stationary or even regress. In the region of the larger flattened forms of sub-capsular traumatic opacities the total depth of the cortex may be thinned considerably (lens atrophy).

Davidson has also drawn attention to such forms of lens opacity and has stressed their medicolegal importance. Nothing is known concerning the pathogenesis of these small traumatic flakelike or punctate subepithelial opacities. They are very similar to those seen following acute attacks of glaucoma. In the latter case actual ruptures of the capsule would be doubtful. In both traumatic injury and glaucoma localized damage to the capsular epithelium resulting from suddenly increased intra-ocular pressure seems a more probable explanation of their origin.

EARLY AND LATE TRAUMATIC ROSETTE-SHAPED OPACITY

Probably the most distinctive form of cataract associated with trauma is the so-called "rosette-shaped" opacity. This type of lens opacity is seen in both nonperforating (contusion) and perforating (penetrating) injuries of the globe. Rosette-shaped opacities associated with perforating and penetrating injuries will be considered under the heading of perforating injuries to the lens. Those seen immediately or shortly after injury are located just below the anterior or posterior capsule (or both) and are known as the fresh type (Fig. 436). The late or delayed traumatic rosette cataract (*remotio cataractae traumaticae*) frequently discovered years after an injury, is situated deeper in the cortex or adult nucleus, and is separated from the capsule by lucid lens substance. It is believed that in these cases the damage occurs to the subcapsular fibers at the time of injury, but the changes are not noticed until years later when they become more opaque. At this time, owing to backward displacement by new clear lens fibers, the rosette is seen to be located deeper within the lens. If the injury occurred early in life, it may be expected that the opacity would be deeper than if the trauma were sustained later.

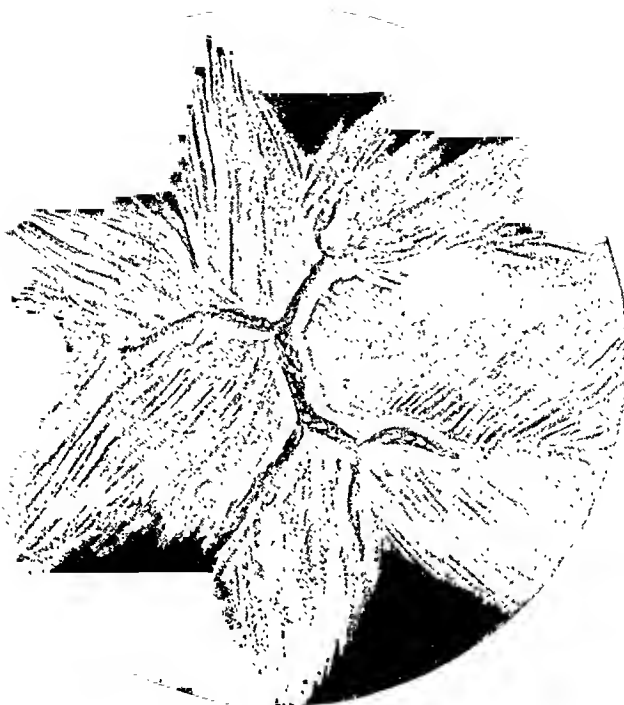


FIG. 436. Traumatic rosette opacity.

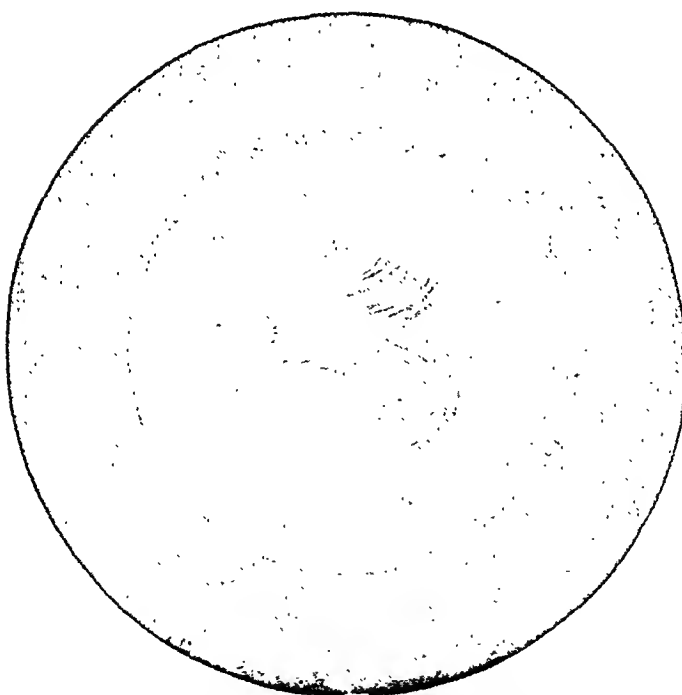


FIG. 437. Traumatic rosette opacity seen in a 5-year-old child. Located deeply about the fetal nucleus. Injury was sustained at the age of 2.

Thus in the former case the rosette may be found behind the adult nuclear zone (assuming that the adult nuclear zone becomes definitely delineated anywhere from the middle to the end of the second decade) (Fig. 437), or if the formation of the adult nuclear band is interfered with, it may be seen in a layer deeper than the surface of the adult nucleus in the fellow healthy eye. But if the injury occurs during or after this period, one should expect to find the opacity at the site of the adult nucleus or in the cortex in front of it. As evidence contributed to the thesis that the original location of a deep rosette (seen years after an injury) is subcapsular, Vogt suggested, the fact (1) that rosettes which develop immediately after injuries generally are found to be subcapsular; (2) that in some cases a few residua of the opacity remain adherent to the capsule and behave like capsular cataracts; and (3) that the rosette form corresponds to the arrangement of the fibers in the subcapsular zone. From his observations and from those of others it appears that the late rosette (almost without exception) is a sign of contusion, which in most instances occurred during early life. Frequently the injury itself may have been forgotten by the patient.

If it were possible to account for all the variables, knowing the time of the injury, it might be possible in some of these cases to determine the growth rate of the lens in the period following the injury.*

In fresh cases the intensity of the opacity may vary from a slight haze to a dense, white, shell-like layer, the variation probably depending on the degree of trauma and the time of observation. In the first instance the lesion consists of a thin subcapsular layer of

* Koby states: "In judging the depth of a rosette we must always bear in mind the possibility of an arrest of development and similarly of an atrophy of the lens. When measuring the total thickness of an injured lens and comparing it with that of the lens of the other eye, we must naturally take care that all conditions of measurements are the same." However, Vogt has shown that in cases where the injuries occurred early, i. e., in the first or second decade, the lucid layer (between the capsule and the rosette) was as much as one-sixth to one-seventh of the total lens thickness. In injuries sustained during the middle of the third decade, this distance was considerably less. Bellows, citing Nordmann, states: "that given a fresh case of traumatic rosette cataract, its future position may be predicted. Thus at the end of one and one-half years it will be deeper than the zone of discontinuity (line of disjunction [M.L.B.]) and in three or four years the opacity will lie in the region of the adult nucleus. He (Nordmann) points out that in younger individuals the apparent inward migration of the opacity is more rapid than in older persons."

fine droplets, lying between the radiating lens fibers. Reflection of light from these droplets results in a grayish opacity the periphery of which has a feathery appearance. The design of the fibers (feathery parallel lines) is thus seen better toward the periphery than adjacent to the usually darker appearing sutures. Vogt made an important differentiation between the appearance of the recent rosette and that of the late rosette. In the latter the suture ends mark the beginning of the dark incisures between the opaque "petals" of the rosette, so that the expansions or rays of the rosette reach much farther peripherally than the ends of the sutures (Plate LXXV, figs. 2, 3). In the former (especially in recent posterior subcapsular rosettes) the rays follow the direction of the sutures, and the areas between the sutures remain clear to form the incisures.

The rosette shape of the opacity results from the suture design "which the opacity renders clearly conspicuous." A "fresh" rosette or opacity, when it is of minimal severity (reminiscent of simple edema) has been known to clear entirely within days or weeks and leave no trace. This is especially true of certain of the posterior rosettes seen after perforating injuries (page 1267). As a rule traumatic rosettes are stationary in so far as their size and density is concerned and if not too opaque (which is commonly the case), they may not cause progressive loss of vision. But as both Davidson and Vogt have pointed out, their presence may later provoke early development of senile opacities. However, in more severe injuries irreversible reactions occur, and a permanent rosette figure may result. These likewise may vary from white opaque figures with feathery ends to those consisting of a solid, thin porcelain-like layer without any apparent internal fiber design.

Although both anterior and posterior rosettes have been described in cases of recent contusion, it appears from the cases described in the literature that the anterior one predominates. However, in my opinion, in cases of perforating injury of the lens the posterior rosette is more apt to occur. Not infrequently, when both anterior and posterior forms are present, or even in the absence of the latter, extensions lying in the same plane may peripherally outline the

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"equator" of the involved layer. In this way they may connect the anterior rosette with a posterior one or if the latter is not present, it may just extend backward for a short distance into a corresponding layer of the posterior parts of the lens. In several cases reported by Vogt and by Haldiman,⁴⁶⁹ there was a second opacity located peripherally to, and a little in front of the main rosette. Vogt does not consider the presence of the second opacity a rarity in contusion cataract. This second, anterior opacity frequently extends to the equator where it may meet an opaque line running concentrically with the equator. Outside this, smaller discrete, dustlike opacities may occur.*

The rosette figure is not complete in every case since at times only one or two "petals" may be visible, and these probably at the site where the greatest part of the force was transmitted. After especially severe trauma thin, dense shell-like opacities without rosette formation may form and like delayed or late rosettes may be found displaced deeper into the lens after the passage of years. In these cases it is not unusual to find a decrease in total sagittal thickness of the lens.

From the standpoint of differential diagnosis there should be little difficulty in diagnosing the fresh traumatic rosette opacity. With the history of a recent contusion, the finding of a subcapsular rosette figure together with other obvious signs of trauma, i.e., ruptures of the iris sphincter or iris tissue, hyphemia or its residue of pigment, one-sided mydriasis, etc., substantiate the diagnosis. However, delayed rosette opacity may have to be differentiated from complicated cataract, saucer cataract (cupuliform), and a type of senile cortical

* The exact time of formation of such extrarosette opacities is not known but Vogt suggests three possibilities, first that they arise simultaneously but independently, the anterior one subcapsularly and the deeper one in the first zone of discontinuity and that later both were pushed back by new fibers. Second, the deeper of the two located subcapsularly could develop first and be pushed deeper before the more anterior one developed, and finally, the original opacity could be separated into two layers by the ingrowth of new fibers. According to Haldiman, traumatic rosette cataract was observed in several cases immediately following contusion. Two layers of opacity were seen simultaneously: the first subcapsular was gray and the second located behind it in the region of the anterior disjunction line or at the level of the adult nucleus band was more yellowish. When sufficiently developed both showed a leaflike structure. In the course of a few months both were displaced backward, the deeper opacity became more distinct, while the superficial one became partially resorbed.

cataract the radiating structure of which (Handmann) may resemble a rosette. Posterior complicated cataract (secondary to intra-ocular disease) (page 1169) is characterized by a polychromatic iridescence in the early stages and later by the "breadcrumb-like" appearance of the opacities. The opacities are not limited to a single zone of discontinuity but tend to progress from the posterior subcapsular region anteriorly toward the posterior adult nucleus as well as in a radiating manner. Posterior saucer cataract (composed of a vacuolated material) is limited to the posterior subcapsular zone and usually has a yellowish reflex. Its frontal boundary always is sharply demarcated from the lens substance in front of it. In neither of the latter are the feathery fiber design seen nor are the opacities pushed toward the center of the lens by clear lens fibers. In certain cases, especially in their early stages, anterior saucer and anterior complicated cataract may morphologically in a gross way resemble a recent traumatic rosette. The former are directly subcapsular and may have a thin radiating character but they do not have the characteristic fiber design as described above. Unlike the late traumatic rosette they are not displaced deeper with time. On many occasions the author has seen thin radiating senile cortical opacities (Handmann) which by diffuse illumination might be mistaken for a rosette. In these cases the optic section will reveal that the opacities result from typical senile and presenile lens alterations (water slits and laminary separation, etc.) and that they differ from the typical "petal" shaped opacities of the traumatic rosette, the ends of which, although feathered, usually have a regular convex arcuate border.

TRAUMATIC LAMELLAR (ZONULAR) CATARACT

The uniocular formation of typical lamellar or zonular cataract (page 1071) following contusion has been reported by several authors. A review of the literature pertaining to this subject was made by Walter (1917)⁶⁷¹ and by Law (1932).⁵¹⁸ The outstanding points noted by Law were that (1) a great majority occurred before 12 years of age (in 22 cases only 2 occurred after the age of 20 years); (2) in a high percentage the injury sustained was of the nonpene-

trating type (contusion) (Plate LXXV, fig. 6); and (3) of the latter as many as half the patients had subluxated or luxated lens. Zonular opacity of the lens develops from other than hereditary causes, e.g., tetany cataract. Evidently whatever the cause, e.g., toxins or increased permeability of the capsule (in injuries — damage to the epithelium or minute ruptures), the time and duration of the insult is all important. In the young, it is conceivable that when the time of action of the disturbing factor is limited to a short period, subsequent new lens fibers could be laid down in a normal way so that in the end a circumscribed perinuclear layer of opacity separated from the capsule by a lucid layer would be found.* In such cases, if the injury occurred early (before the adult nucleus is definitely delineated) the opacity would surround the outer embryonal or fetal nucleus but if it occurred somewhat later (after adolescence) the adult nucleus itself might be affected. As already noted, in hereditary zonular cataract or in those resulting from exogenous causes (tetany) the opacity may be complete or partial. Riders may or may not be present. I recently observed a case (a perforating injury with marked hyphemia in a 22-year-old man) in which three weeks later, in the operative coloboma, a double layer of typical linear opacities were seen outlining the equator of the adult nucleus. Slight iridonesis suggested that some degree of subluxation was present. Dilatation of the pupil of the fellow healthy eye revealed no coronary or other opacities. The opacities began in an area corresponding to the peripheral one-third of the anterior surface of the adult nucleus and continued around the equator to about the same area on the surface of the posterior adult nucleus — an example of a partial or incomplete zonular opacity. These opacities had a somewhat yellowish

* According to Law: "Such an event could result from a temporary alteration in capsular permeability caused by a severance of the physiological connection between the capsule and the lens, with a consequent alteration in its osmotic properties, or—alternatively and rarely—by a perforating capsular injury which subsequently healed. In each of these cases, degenerative changes would occur in the periphery of the lens as then constituted, if an approximation to natural conditions was restored without delay, the nutrition of the central lens fibers would not be disturbed and the fibers laid down subsequently would again be transparent. According to the severity or duration of the alteration in physical properties of the capsule so would the transparency of the central lens or the younger peripheral fibers be affected to a greater or lesser degree."

tinge. It is of interest that in many of the cases of traumatic zonular cataract varying degrees of subluxation and luxation of the lens have been noted. The frequency with which opacities appear in anteriorly displaced lenses has already been touched upon. The tearing of the zonular attachments in itself may play a role. As Vogt pointed out, atropine mydriasis should be used with caution after ocular injury (see footnote, page 1259) because with relaxation of the zonule, the capsular tension increases and small wounds in it may consequently be caused to gape, thus not only interfering with healing but also permitting greater access of aqueous into the lens substance.

PRESENILE AND SENILE-LIKE LENS OPACITIES INDUCED BY CONTUSION

That lens opacities, morphologically similar to the common varieties ordinarily known as presenile or senile lens alterations, may be initiated or intensified by contusion has been attested to by the findings of many observers. Among these changes are coronary and cerulean (page 1071) opacities, zonular cataract, punctate cortical opacities, water slits and spokes, and nuclear cataract (Fig. 464). Vogt reported on a case in which an explosion twelve years before affected the left eye. The cornea was not injured. In addition to several oval opacities axially he found typical coronary and cerulean opacities in the periphery of the anterior and posterior cortex. These consisted of typical clubs and disks. More axially there were blue and brown disks and larger lancet-like streaks (frequently seen in cerulean cataract). The lens sagittally was thinner than that of the unaffected right eye. However, when the pupil of the right eye was widely dilated, it was possible to make out faint traces of coronary opacities. He concluded that in this case the severe trauma intensified the antecedent cataracta coronaria and cerulea. He also cites other cases where pre-existing coronary opacities increased in number and size following trauma. In this connection, Vogt reminds us of other senile conditions that may also appear when provoked by exogenous causes, e.g., arcus senilis and corneal lines (Hudson-Stahli).

I have several times noted the presence of punctate and dustlike opacities, especially in the peripheral cortex, following contusion. These are similar to those described as senile punctate opacities (page 1123). Such opacities were particularly marked in a case in which the lens was subluxated upward, permitting a better view of the lower equatorial regions of the lens. In many of these cases the fact that the punctate spots were either absent or at best sparse, in the unaffected eye, indicated their connection to the injury.

TRAUMATIC CATARACT ASSOCIATED WITH RUPTURE OF THE LENS CAPSULE

Rupture of the lens capsule in either its anterior or posterior parts (*Perforatio Lentis Traumatica*) invariably results in cataract formation. Although tearing of the capsule may follow contusion even in the absence of any demonstrable rupture of the outer tunics, usually it is associated with perforating or penetrating lesions. The biomicroscope is of inestimable value in detecting faint, perforating corneal wounds (especially those at the limbus), small holes in the iris, scleral or conjunctival hemorrhages, and conjunctivo-scleral scars (especially pigmented) — all indicative of perforation. These are frequently overlooked by cruder methods of examination. Confirmation of the perforating nature of the injury may be obtained by the ophthalmoscopic presence of chorioretinal scars in the fundus behind the site of the above mentioned lesions, i.e., along the track of the penetrating missile. But it must be admitted that in certain instances very small penetrating foreign bodies may leave no external trace of their passage into the eyeball. Hence, it is axiomatic that when one is confronted by an inexplicable unilateral lens opacity, radiographic examination should be made for the presence or absence of a foreign body.

The opacification of the lens following rupture of the capsule probably is due to direct injury of the fibers themselves and the action of the aqueous upon them. Hence it is conceivable that with rapid sealing off or closure of a capsular wound after a penetrating injury, a localized stationary lens opacity could result. Such a con-

dition is seen frequently following penetrating injuries (by sharp instruments or foreign bodies), especially when they have entered the peripheral parts of the lens. On the other hand, a slight rupture of the capsule (especially in the axial regions) may be followed within hours by rapid intumescence, leading to total cataract.*

With large capsule rents the lens substance rapidly swells to a high degree so that obliquely placed fibers are often seen protruding either into the anterior chamber or into the vitreous. In the young, depending on the circumstances, partial to complete absorption of lens substance follows so that in the end conditions varying from secondary cataract to aphakia may occur (see Fig 453). In older individuals, the inabsorbable sclerosed nucleus when exposed provokes a foreign body reaction with dire results. Even in the absence of infection disintegrating lens matter may act as an allergen, inducing complications (uveitis [phaco-anaphylactica] and secondary glaucoma). As in severe contusions, traumatic perforation of the lens capsule may also result occasionally in the appearance of either partial or complete anterior or posterior rosette opacities, the posterior probably being more common (Figs. 441 A, B; 442; 443). The rosette may be found at the first examination after the injury

* In his remarks about the behavior of capsular wounds, Vogt stated that it is well known that circumscribed wounds of the lens behave differently from case to case and that their course is unpredictable. This also applies to "needling" operations. In one instance a small-sized opening may lead to a total cataract and in another it may close quickly. Likewise in lenses in which the capsular opening is large and even when perforation of the lens substance has occurred, a small localized opacity may result. This also applies to luxated lenses, especially those where the displacement occurs spontaneously. Surgeons know how difficult it is to cause these to mature by needling. In several cases, even after two needlings Vogt succeeded only in obtaining a limited opacity at the site of the capsular tear. Perhaps the surrounding vitreous played a protective role in some of these cases but it was difficult to obtain opacification of the lens even when there was no vitreous in the neighborhood of the wound. He concluded that the cause of this special behavior in luxated lenses is to be found in the fact that owing to the lack of zonular and capsular tension, there is less tendency for capsular wounds to gape. This also holds true for animals (rabbits and lower vertebrates) in whom, as was previously pointed out by Knapp, it is difficult to cause opacification of the lens by needling. Owing to the lower power of accommodation in these animals their capsules are less taut and stretched than is the case in humans. Reasoning from these observations, Vogt cautioned that the use of atropine (except in the presence of infectious inflammation) immediately after capsular ruptures should be avoided since with paralysis of accommodation there is an increase in stretching or tension of the lens capsule. Hence, when possible, relaxation of the zonule with miotics may be useful. For the same reason, when the capsule wound is located behind the iris, it might be dangerous to atropinize the eye at an early stage when the iris may furnish a useful and protective exudate. Iris traction (by atropine) conceivably might tear open the freshly adherent capsular wound.

and may either retrogress (edema ?) or, when irreversible damage to the lens fibers has occurred, may become more opaque and consequently stationary. As a matter of fact, the lens opacities following the penetration of foreign bodies, except those marked by a complicating siderosis, do not differ essentially from those described as contusion opacities, with the possible exception of the senile type of contusion opacities (page 1257). The partial lamellar type, in which the single-layered opacity extends around and accents the equator of the layer in which it lies (like zonular opacities), is not uncommon (Plate LXXI, fig. 6). This suggests that the penetrating foreign body may produce the effect of contusion.

At this place it is appropriate to mention the danger of inducing traumatic cataract (rupture of the capsule) at the time of operative intervention for glaucoma. Kirby⁴⁹⁴ pointed out that Wheeler warned about this accident (improper keratome incision; attempting to grasp the iris too close to its pupillary border, etc.). In the presence of high intra-ocular pressure, it is possible that owing to the sudden decompression following upon opening of the anterior chamber, that rupture of the posterior capsule may occur.

Davidson³⁹⁴ states: "(1) A survey of 62 lens lesions, primarily opacities, observed in cases of eye perforation by intra-ocular foreign bodies and in eye rupture without retention of foreign body, indicates that the majority are the result of lens contusion and morphologically belong to the type of contusion-lens opacities previously studied. Posterior feathery star-shaped opacities are evidently rare.

"(2) Capsule perforation, lens penetration, or its double perforation occur in less than a third of the cases that do not lead to immediate cataract.

"(3) The retrospective diagnosis of the eye perforation or rupture origin of a lens opacity is sometimes made difficult because of the eventual blurring of the perforation or rupture character of a corneal scar after many years and particularly because of the difficulty in diagnosing an older partial limbus and scleral perforation or rupture.

"(4) The rate of recession into the depth of the lens of originally

subcapsular opacities is found to vary . . . just as in the study of pure contusion lens opacities. The factors affecting the rate seem to be: capsule lesions, which tend to retard it; varying depth of lens penetration, which would tend to accelerate it; hypertension [increased intra-ocular pressure, M. L. B.] and siderosis, which tend to retard it; and hypotension [reduced intra-ocular pressure, M. L. B.], which favors it.

“(5) As for the evolution of the lens lesions in this mild variety of cases, the end results are satisfactory in the majority of cases, but prognosis should be more guarded in the individual after he is 30 years old. Deterioration should be watched for in the fifth decade when apparently most deteriorations occur.”

From the standpoint of the biomicroscopic appearance the following changes in perforating injuries of the lens will be considered:* (1) visible capsular wounds and scars; (2) posterior subcapsular rosette cataract and its resorption (Fuchs, Avizons, Vogt); (3) localized (stationary) opacities (cataracta traumatica circumscripta); (4) intumescent (total) traumatic cataract; (5) traumatic folds of the capsule and (6) changes in sagittal lens thickness (post-traumatic atrophy of the lens — Vogt).

VISIBLE CAPSULAR WOUNDS AND SCARS

The biomicroscope permits the inspection of the anterior and posterior parts of the lens capsule over an area limited only by the size of the pupillary opening. The equatorial areas ordinarily present difficulties in visualization, except in the presence of an iris coloboma, ectopia lentis, or following iridodialysis. But even in these cases damage to the capsule at the equator is not easily demonstrable. In many instances sharp delineation of the capsule stripe by means of the optic section may not be possible immediately after the injury owing to the corneal changes (edema), and to increased turbidity of the aqueous either from exudate or blood cells. When the capsule stripe can be clearly seen, any break in its continuity will be appar-

* It should be remembered that like traumatic injuries in general, no two are morphologically alike and hence one can only describe them generally.

ent as the beam is passed over its surface. Capsular wounds tend to gape somewhat and the edges tend to roll outward. Individual opaque fibers or masses of opaque lens matter often may be observed to protrude mushroom-like into the anterior chamber. After a time, within the unabsorbed protruding opaque mass, fine chalky crumblike granules and dots form. When these are seen in a capsular wound, it is an indication that the lens masses are no longer fresh and that the condition has existed for some time.

Immediately following the injury, the edges of the wound become more opaque, often resulting in the formation of an oval ring or slit of varying size. Protrusion of opaque fibers through this opening is not seen in every case. A halo-like second ring, grayish and opaque, may be seen to surround the inner ring. There may or may not be any bulging of the lens within the ring. With high magnification it may be seen that the grayish halo (surrounding the capsular wound) which at times contains pigment granules has the character of an exudate, adherent to the capsule. Evidently a gap in the capsule may become covered over with a thin layer of fibrin. Vogt described such a case in which two years after injury by a perforating foreign body a spontaneous rupture occurred within a well-formed ring, so that at this late date opaque lens masses suddenly began to protrude into the anterior chamber. When a perforating object passes through the iris (leaving a hole) near the pupillary margin, an iridic adhesion to the capsular wound may occur because of exudation. This may serve to plug the capsular wound, thus limiting the extension of the cortical opacification.

It should be noted that following perforation of the lens by small foreign bodies, opacities of the anterior capsule may not always be found biomicroscopically. In his series of 62 cases, Davidson was able to demonstrate capsular injuries in only twelve cases. In six of these there was double lens perforation with a demonstrable tract in the lens. In one case the capsular lesion was seen at a distance from the end of the tract similar to the cases reported by Meesmann. Considering the fact that the direction of most perforating foreign bodies is oblique it may be that the lens often deflects them or else



FIG. 438. Traumatic cataract. Dense capsular scar. Folds in capsule. Anterior line of disjunction is not seen in the region of the scar. (After Vogt.)



FIG. 439. Capsular scar. Note fiber design at its edges; also capsular fold running through it. (After Duverger and Velter.)

their entrance is not demonstrable. In the case of the former the resulting opacities may be the expression of a contusion. Davidson noted that double lens perforation and anterior capsule perforation were conspicuously more frequent in the older age groups — a condition possibly accounted for by capsular friability.

With healing, a dense scarlike opacity may result, and characteristically the anterior line of disjunction is usually not distinguishable within it (Fig. 438). At the margin of the opacity the fiber design may be visible as radiating white lines (Fig. 439). The presence of pigment at the site of a capsular scar should always be looked upon with suspicion. The force exerted by the penetrating objects may cause the cornea to be momentarily pressed backward against the iris; this in turn strikes the lens so that pigment may become adherent to the capsule or forced into the lens at the site of the capsular wound. It is possible that when a penetrating object passes through the iris, some of its pigment may be carried along with it. In the neighborhood of capsular opacities it is not unusual in the zones of specular reflection (mirror region) to observe iridescence. This phenomenon is probably due to interference effects.

As a consequence of the formation of a capsular scar, traction folds may result. They may radiate fanlike or may extend irregularly. Irregularities in the curvature of traction folds are frequent. In the latter case the narrow beam may show troughlike depressions. Within the center of old perforation scars of the anterior capsule, occasionally a "blister-like" lens spherule may appear. These resemble dark structureless giant subcapsular vacuoles but differ in the fact that they protrude from the opaque surroundings (Fig. 440 A, B). Although they are dark in direct focal light, they appear to have a delicate whitish envelop and hence are unlike the so-called "Kugeln" or grapelike cysts (Elschnig, Hirschberg) usually associated with secondary cataract or membranes. Because of the opaqueness of the surrounding scar it may be difficult to observe them by retroillumination. Such bodies may represent a cystic degeneration of the capsule or, as suggested by Vogt, "an epithelial product." They have not been seen on the posterior capsule.

In a 7-year-old child, who suffered a perforating injury with prolapse of the iris (which was excised) Vogt found an aggregation of clear subcapsular spheres at the equator, at a site opposite to the

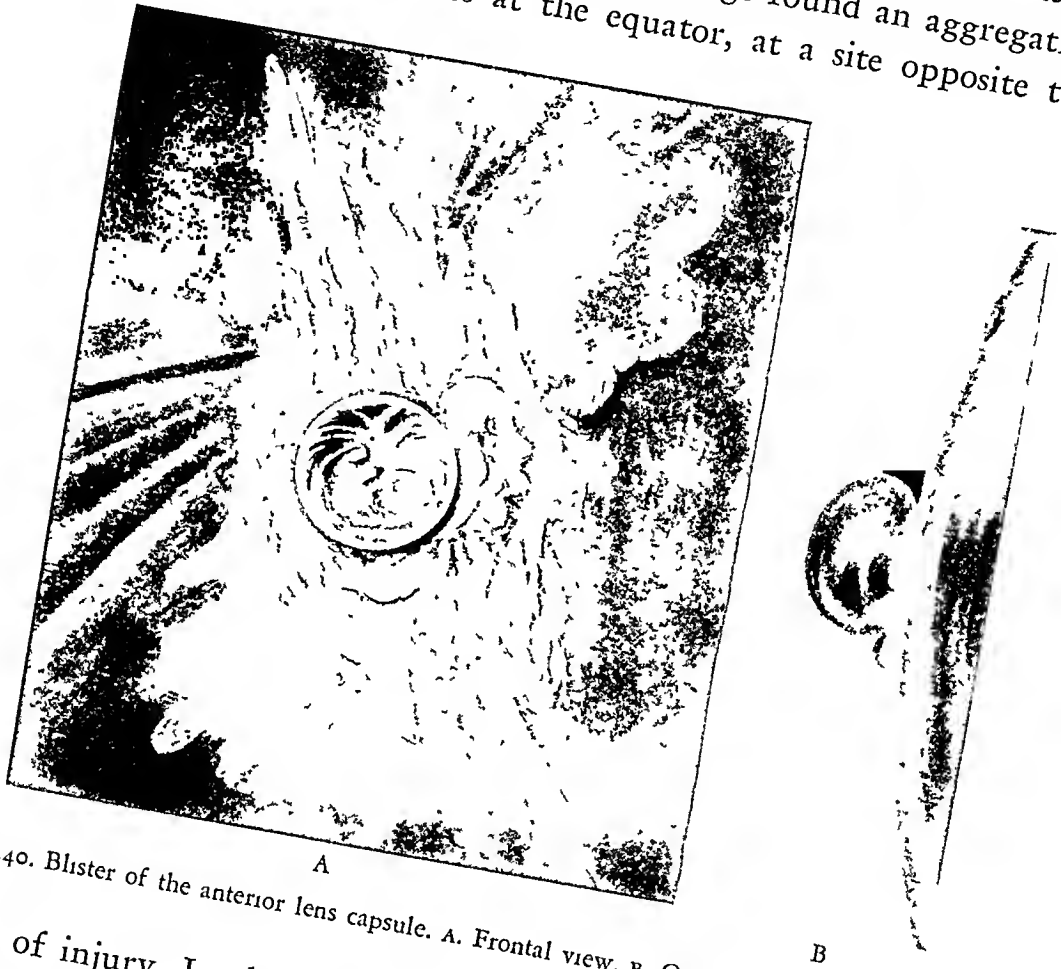


FIG 440. Blister of the anterior lens capsule. A. Frontal view. B. Optic section (After Vogt.)

place of injury. In the upper region of the lens near the equator there were some elongated subcapsular vacuolar clublike structures that were similar in shape to the opacities seen in coronary cataract. He suggests that the accumulation of fluid in this form might explain the origin of coronary opacities, in that the latter may be a further extension of similar fluid collections. In this region (equatorial) there are no sutures and consequently the lens fibers have a more decided radial course.

Injuries in which the penetrating object has traversed the entire lens substance and has damaged or perforated the posterior capsule, will naturally cause changes in this part of the lens. As in contusions, rupture of the posterior capsule may occur in other sites inde-

A



B

FIG. 441. Anterior rosette seen several years following a perforating injury. Site of perforation near equator is not shown. A. Diffuse view. B. Optic section.

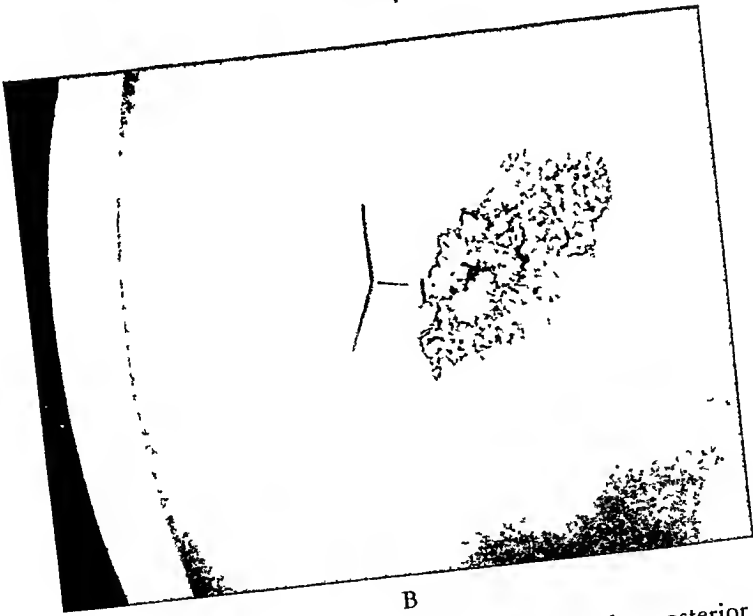
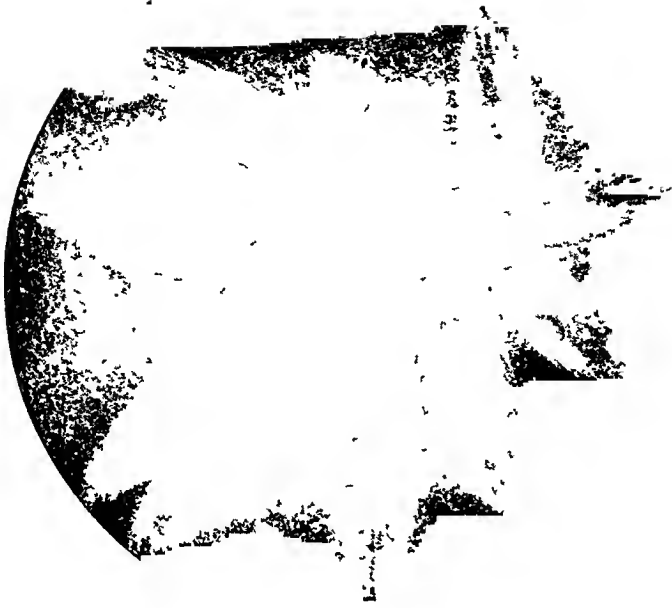
pendently of the actual passage of perforating objects through it. This may happen also in cases where the penetrating object does not reach the posterior capsule. As a result of compression from the force of the injury (as explained by Frankel) in the contusion syndrome there is an increase in the equatorial diameter of the lens. This may by itself rupture either the zonule, the posterior capsule (which owing to its thinness is more liable to rupture than the anterior capsule), or both.

Localized posterior subcapsular crumblike opacities may be found which resemble posterior complicated cataract (page 1168) and may lie in front of scars of the posterior capsule. In adults, owing to increase of yellow tones in the posterior parts of the lens the crumblike opacities may have a yellowish color in contrast to the whitish aspect of the more anterior parts of the opacity. Following perforation of the posterior capsule, opaque lens masses may be forced backwards into the vitreous. These masses will oscillate within the vitreous with movements of the eyeball. As a rule, the exit wound of the posterior capsule tends to be larger than that of the entrance wound of the anterior capsule. This is to be expected because as the foreign body passes through the lens a larger area of displaced tissue is forced backward. As in the case of myelin droplets the light reflected back from the fundus may cause the edges of a posterior capsular wound to glow with a reddish color. Upon healing, the optic section frequently shows that the scar in the posterior capsule bulges concavely backward.

POSTERIOR SUBCAPSULAR ROSETTE CATARACT AND ITS RESORPTION

As mentioned previously in the section on contusion cataract, rosette-shaped opacities also may develop following perforating lens injuries (Figs. 441 A, B; 442 A; 443). They usually occur in the cases in which the opening rent in the capsule has been small and in which owing to capsular closure, complete opacification of the lens (intumescence) does not obscure this delicate change. Consequently most of the cases that show such capsular closures are instances of damage caused by the penetration of pointed instruments (needles)

A



B

FIG. 442. A. Posterior rosette after perforating injury. B. Irregular posterior cortical opacities seen to one side of the optic section by retro-illumination. Perforating injury.

or small foreign bodies. Within a few hours or during the first few days after the injury biomicroscopic examination, especially in the posterior parts of the lens, may reveal the presence of a rosette

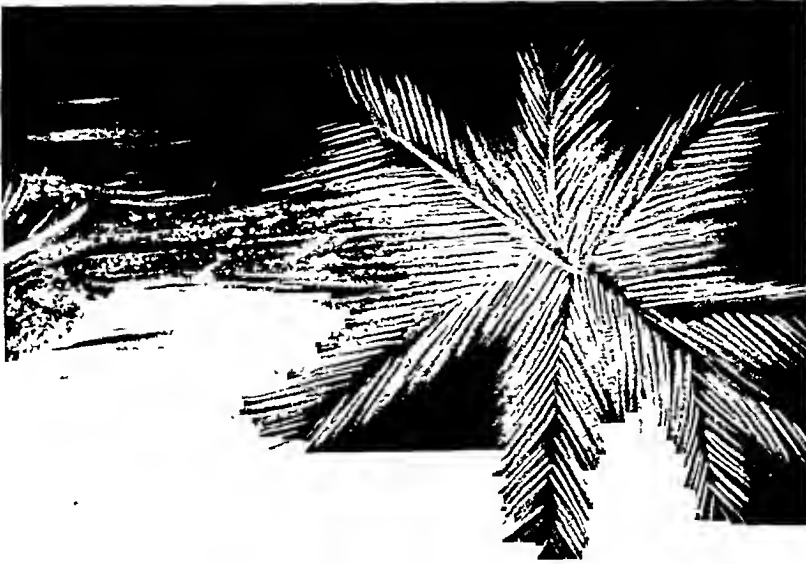


FIG. 443. Perforating injury. Posterior rosette.

opacity. It is located just in front of the posterior capsule (axially), and its structure sharply outlines the suture and fiber design. The rosette tends to lie more or less in one plane and is relatively of little thickness. The sutures appear dark and extending from them are feathery grayish to yellowish lines (depending on the color of the posterior cortex). Each division or "petal" of the rosette can be compared to a feather in which the central spine or suture is dark, thus differing somewhat from the contusion rosette where the end of the suture marks the beginning of the incisura. In direct focal illumination the surface of the rosette may glitter, somewhat resembling the sheen from a thin layer of crystallized boric acid (Vogt). Transillumination by means of the ophthalmoscope reveals a vacuolated structure, which gives the impression of a figure formed by a layer of subcapsular fluid or exudate outlining this fiber design (Fig. 443). According to Vogt, the sutures are a *locus minoris resistentiae*, permitting the entrance of fluid that reaches the fibers. This observation may explain the physiologic manner

in which the nourishing fluids get to the fibers. The suture planes extend through the whole depth of the lens substance and hence represent the only way in which fluids could gain access to individual fibers. The fact that this type of opacity can resorb within a short period after its appearance (a fact first noted by Fuchs) would indicate that the fibers in themselves are not irreversibly damaged. Avizonis³⁵⁰ reported a case in which a posterior contusion rosette cleared entirely within a month. In one of Vogt's cases the original size of the rosette was 45 mm. in diameter. Within a month it shrank to 0.75 mm. in diameter so that with the ophthalmoscope only a few faint dots remained visible. The vision during this time improved from 6/18 to 6/8. In these cases when a penetrating object passes through the lens, an opaque tract extending from a wound in the anterior capsule to a posterior rosette figure will be seen (Fig. 443) and if the posterior capsule is injured, there may be an opacity in it either in connection with the rosette or just behind it. However, a posterior rosette figure can form in the axial region even if the injury does not extend through the entire lens thickness and also when the direction of the entrance wound is not axial, e.g., at the equator. According to Wagenmann,⁶⁶⁷ even when the injury is localized to the anterior part of the lens fluid very quickly may make its way posteriorly by following the equatorial curve of the involved plane so that an opacity remote from the injured site may occur. This again would emphasize the concentric lamellar structure of the lens — an architecture seen physiologically in the behavior of the zones of discontinuity.

LOCALIZED TRAUMATIC STATIONARY OPACITIES

In addition to the rosette opacity just described which has a predilection for a special locality in the lens and the form of which follows the morphologic structure of the lens, perforating lens injuries may cause permanent localized and stationary cataracts (*Cataracta Traumatica Conscripta*). These consist of irregular opacities, varying in size and shape from small dots or lines to large cloudlike masses, limited to the injured area (Fig. 442 B). In the

periphery, flattened thin opacities having a pyramidal (cuneiform) or semilunar shape are not uncommon. Many extend posteriorly by passing around the equatorial region of the involved plane similar to lamellar opacities.

The track of a penetrating injury may be marked by a linear opacity which consists of streaky lines or dots or may have a solid spear-like character. With rapid closure of the capsule this may remain as permanent unchanging evidence of a past perforating wound. When peripherally located it may be necessary to dilate the pupil to see such opacities. After the passage of a foreign body through the equatorial region of the lens it is not unusual to see more or less localized irregular masses of opacity limited to this area. They are cloudlike and not sharply delimited, show little tendency to progression and may not markedly impair vision.

Several years ago I saw the case of a 27-year-old man in whom a tract of scar was seen traversing the corneal thickness of the left eye at about 5 o'clock near the limbus. Just behind this there was a small hole in the iris, which only became apparent by retro- and transscleral illumination. It was only upon maximal dilatation of the pupil that a large fluffy opaque cataract, involving the entire thickness of the equatorial region, could be seen (Plate LXXI, fig. 5). The patient came to the clinic for a refraction unaware of any previous trouble and it was only on questioning his parents that a history of a long-forgotten injury in early childhood was obtained. At the age of three, this eye had been perforated by a hat pin while the boy was playing with his older sister. I have seen several similar lens opacities following perforating foreign bodies that passed through the periphery of the lens. These opacities may show no vacuoles, water slits or fiber design, and usually they are not limited to any one zone of discontinuity. Similar irregular opacities in the equatorial regions may be seen in cortical senile cataract, but with careful inspection particularly at the axial borders, it will be noticed that they are the extension of either a radial spoke or cuneiform opacity.

INTUMESCENT AND TOTAL TRAUMATIC CATARACT

As a rule, larger capsular rents tend to provoke total cataract because of the greater separation of their edges. However, as previously



FIG. 444. Intumescent cataract. The fibers are separated and twisted.

pointed out there is no definite relationship between the size of the capsular opening and the amount of lenticular opacification which ensues. Openings of the capsule (anterior or posterior) may lead to rapid intumescence and complete opacification. The end result of this process will vary in each individual case, depending on whether or not the capsule rent closes, on the age of the patient, and on the presence of other intra-ocular complications. In the young, partial or complete absorption of the capsular contents may occur, resulting in aphakia. After the third decade, the presence of a ruptured capsule and intumescent lens is more serious, and because of the complications (iritis, glaucoma or even sympathetic ophthalmia), removal of the cataractous lens may become imperative.

Intumescence is manifested biomicroscopically by the separation of the opaque fiber bundles by dark slits or gaps. At times fiber bridges (comparable to twisted and separated wool fibers) cross the

gaps obliquely (Fig. 444). The similarity of this picture to that seen in senile intumescent cortical cataract has been pointed out, and one gets the impression that water imbibition has forcibly torn the fibers apart. In other cases a layer of subcapsular vacuoles may be seen in front of opaque cortical masses. Protrusion of swollen lens fibers out of the capsule and into the anterior chamber not only tends to keep the capsule rent open but also permits the continued lytic action of the aqueous on the fibers.

With closure of the capsular opening (by exudate or adherent iris) the process of intumescence may be localized to a minimal extension of the cortical opacification. In this case the lesion may be limited to some short fine superficial stripes radiating from the capsular scar, showing no further signs of progression. Cases have been reported in which spontaneous reopening of a sealed-off capsular rent occurred years after the original trauma, with consequent extrusion of lens matter into the anterior chamber.

In injuries to the posterior capsule the tendency to intumescence of the cortex is not as rapid or widespread as in the anterior cortex. It may be that the presence of the vitreous in juxtaposition to the posterior capsule may mitigate against the entrance of fluid into the posterior cortex, thus inhibiting rapid opacification. Consequently the resulting lesion will tend to be more localized. This is emphasized by the frequent presence of localized capsular scars and opacities of the posterior cortex.

However, in cases of double perforation by foreign bodies, rupture of the posterior capsule (the exit opening may be larger than that in the anterior capsule) may be followed by the protrusion of the opaque lens masses into the anterior vitreous. Some of these opaque fibers may become detached and may float with the movements of the vitreous, and later, if visibility through the lens permits, may resemble the bodies of asteroid hyalitis.

TRAUMATIC FOLDS OF THE CAPSULE

Folds of both the anterior and posterior lens capsule may follow perforating lens injuries, especially in cases in which traction scars

develop in the capsule. As a rule the folds tend to radiate from the scar. Ruptures of the capsule may also cause folding by kinking, even in the absence of contracting scars. Capsular folds show characteristic double reflexes and are usually pointed at their ends, having a lanceolate appearance (Fig. 439). Those of the posterior capsule can be verified by use of the optic section. Here adherent pigment particles and opaque vitreous strands may be observed. Bright red blood clots, adherent to the folded posterior capsule, may also be seen for months after an injury. Folds of the anterior capsule may occur even without capsular traction scars in cases of contusion cataract which tend to become hypermature (Plate LXXI, figs. 1, 2). After perforating injuries, adhesion of a torn capsule to either the iris or cornea may cause capsular folds in the neighborhood of the adhesion.

CHANGES IN SAGITTAL LENS THICKNESS

In demonstrating changes in the sagittal dimension of the lens, especially reduction in volume that may occur after severe trauma, use of the optic section is essential. As already indicated, such reductions in volume may also occur after severe contusions in which no capsular opening can be found (posttraumatic atrophy of the lens). Following a contusion, Vogt found that the ratio of the sagittal thickness of the injured lens to that of the uninjured lens in the fellow eye was 2:3. In the cortex behind the somewhat flattened capsular stripe which contained spotlike opacities (Fig. 445), there were two well-marked zones of discontinuity. The deeper one fused peripherally with the adult nuclear stripe, which was flattened and very relucant. The peripheral convergence of the zone of discontinuity was probably a result of the disturbance that caused a reduction in lens volume. Contrary to the general occurrence in cases in which the loss of sagittal thickness follows perforation of the lens capsule with consequent shrinking, folding of the capsule was not seen. The cause of the reduction in volume of the lens after the contusion could be explained by resorption of the opaque or damaged fibers and also by a disturbance of growth resulting

from epithelial damage. That thinning of the sagittal lens thickness should occur following perforating injuries is thus to be expected. Usually the thinning is more likely to be found in the neighbor-



FIG. 445. Changes in sagittal lens thickness. (After Vogt.)

hood of the perforation — the place where opacification and destruction of lens substance is greatest. In this region, besides opacification, large subcapsular water-slits may be found. In these cases folds of the capsule, similar to those described in hypermature cataract, are common. Damage to the lens epithelium, especially if the wound is equatorial, would prevent new lens fibers from growing in or at least would impede their full growth. Hence if apposition of lucid young fibers does occur, it will only appear as a thin clear layer and will not add considerably to the total lens thickness.

In contradistinction to thinning of the sagittal thickness of the lens after contusions and perforating injuries, an increase in sagittal lens thickness may be noted in cases of chronic glaucoma. In these cases the sagittal thickness of the nucleus (which is more reluctant than normal) seems wider at the expense of the cortex.

LENS CHANGES FOLLOWING PENETRATING FOREIGN BODIES

The penetration of foreign bodies into the eyeball constitutes a large percentage of industrial ocular injuries. The lens may be directly injured, or changes in it such as complicated cataract may be set up secondarily in consequence of effects of the injury (e.g., inflammation or infection) or by the retention of certain metallic

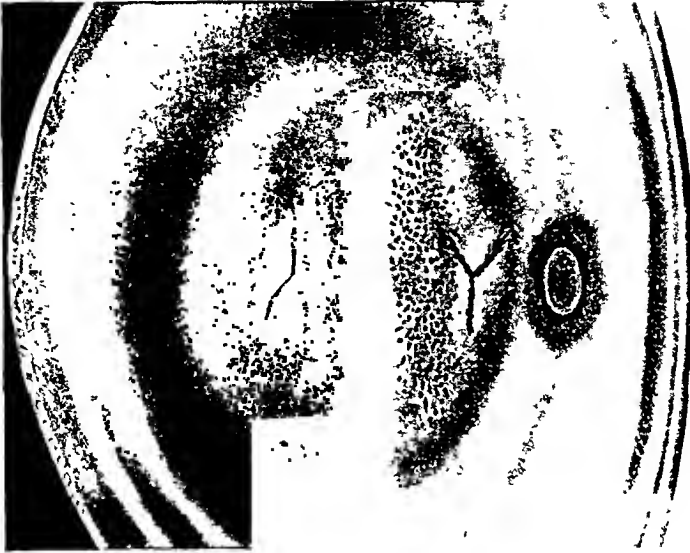


FIG. 446. Foreign body in the lens located near the anterior Y. The opacity of the anterior suture and the punctate opacities are congenital, since they were seen also in the unaffected right eye. (After Duverger and Velter.)

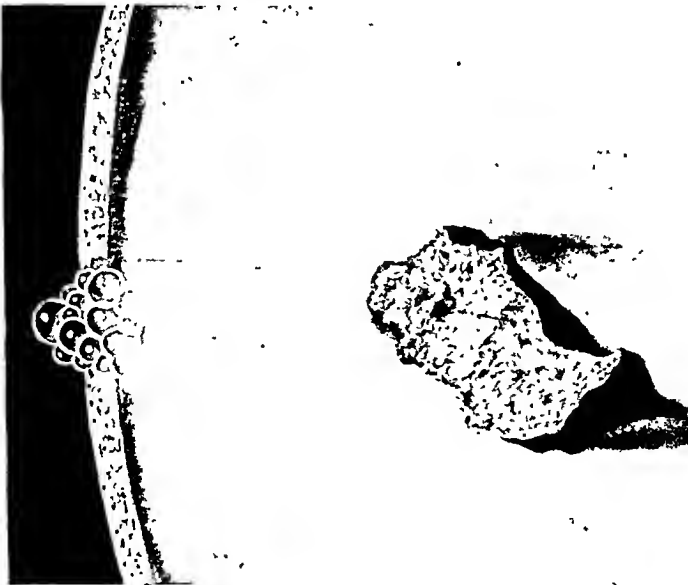


FIG. 447. Foreign body in lens. Note blister-like degeneration of capsular epithelium at the entrance wound in the capsule. Total cataract. (After Vogt.)

foreign bodies within the eye (e.g., iron and copper). In the case of the latter, owing to physicochemical causes, metallic impregnations occur within the lens as well as in other parts of the eye (the epithelial tissues being especially vulnerable). With the exception of iron and copper, it is well known that — excluding the direct local injury to the lens fibers and the effect of open capsular rents (lytic action of the aqueous) — the lens tolerates most foreign substances very well.* The effects of the penetration of such substances differs in no way from those described under penetrating injuries of the lens. In a few cases inert foreign bodies in the lens, even when they have been present for a long time, have been known to produce only a localized opacity without any complicating reaction (Fig. 446). However, in most cases total cataract eventually occurs (Fig. 447). Frequently even in the absence of introduced infection or toxic effects from retained iron or copper the presence of an intra-ocular foreign body results in a plastic iridocyclitis with or without increases of intra-ocular pressure. Such a condition may excite sympathetic ophthalmia. The importance of biomicroscopic examination in suspected cases of intra-ocular foreign body cannot be too strongly emphasized. It may be the only way by which it is possible to detect delicate wound tracts in the cornea or neighboring sclera, iris, and lens. Discoloration of the cornea, iris, the anterior subcapsular regions of the lens, and the vitreous in siderosis, chalcosis, or hydragryosis may only be revealed through the medium of biomicroscopy. Even in the absence of any discernible wound tract, the appearance of an otherwise inexplicable monocular cataract always warrants radiographic investigation. Foreign bodies entering the lens through the pupil are less liable to cause an immediate iritis or hyphemia than those in which perforation of the iris has occurred.

* In World War II as a consequence of aerial flak, secondary fragments of plastic material from the windshields or turret blisters were not infrequently seen in wounded personnel. With the biomicroscope small slivers of slightly refractile homogeneous plastic surrounded by a faint area of opaque lens matter were seen embedded intralenticularly. These in themselves appeared to be well tolerated within the lens. Also as a result of plastic mine explosions, the green plastic material employed in the construction of the German mine was discovered embedded in the lens as well as elsewhere in the eye.

PLATE LXXVI

FIG. 1. Siderosis lentis. Diffuse illumination. Total cataract. Note the radial disposition of the deposits.

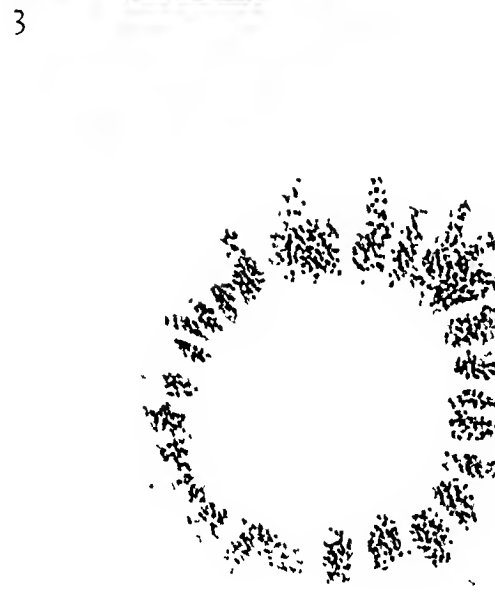
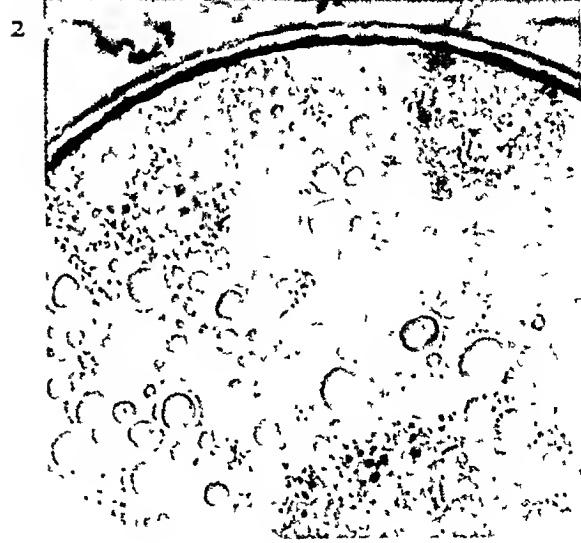
FIG. 2. Same case as shown in Figure 1. High power. Diffuse illumination. Note the groups of small dotlike deposits and also larger areas of stain. The iris is discolored.

FIG. 3. Siderosis lentis. Posterior complicated cataract with polychromatic iridescence. Pigmentation of the vitreus.

FIG. 4. Sunflower cataract in chalcosis lentis.

FIG. 5. Subcapsular location of the deposits in chalcosis lentis.

FIG. 6. Posterior complicated cataract in retinitis pigmentosa associated with unusual pigment deposits on the posterior lens capsule.



SIDEROSIS OF THE LENS (SIDEROSIS LENTIS)

A deposition of "rust" in the lens occurs either when a ferrous particle has perforated or lies within the lens or as part of a general ocular siderosis even in cases where the lens itself was not injured primarily. In the former case one may find localized areas of discoloration in the neighborhood of the particle or splinter, while in the latter, when the metal lies outside the lens, a more characteristic and regular disposition occurs. However, exceptions frequently are seen. As is the case with other perforating foreign bodies of the lens, probably dependent on capsular closure, a localized opacity with varying degrees of neighboring discoloration may occur, or with rapid intumescence a total cataract may follow, this also may show varying degrees of rust deposition.

The more characteristic and regularly disposed type of lens siderosis is usually seen in cases which exhibit involvement of the other ocular tissues (cornea, iris, vitreous and retina). Despite the well-known affinity of the intra-ocular epithelium for ferrous metals, it has been shown by Koepe, Vogt and Meesmann that in siderosis bulbi the stroma of the cornea as well as the vitreous may be involved. In these cases the discoloration in the lens (siderosis) is found with the narrow beam to be located just behind the anterior capsular stripe or in the region that probably corresponds to the location of the capsular epithelium. Morphologically the deposit may be seen in the form of minute little yellow to brownish colored dots or as larger brownish to reddish accumulations (Plate LXXXVI, figs. 1, 2, 3). The smaller dots may be generally distributed in more or less of a single plane over the lens in the periphery as well as the pupillary area. The dots are frequently arranged in groups in such a way that a pattern may form. It appears as if clear (unaffected) irregular channels or "gyrus" spaces form between the groups of dots. In certain areas the number of dots may be more numerous than in others and when this is so the clear channels become narrower or are missing. Except for a slight increase in yellowish color, probably caused by reflection from the underlying stained epithe-

limum, the shagreen of the capsule is not affected. The larger accumulations form well defined rust spots frequently visible to the unaided eye (especially in daylight), being from 0.5 to almost 2 mm. in size. This is especially true when, as is frequently the case, the lens is opaque, the sub-capsular area of cortex being vacuolar, thus producing a striking contrast between the rustlike spots and the grayish background. In distinction to the more general arrangement of the smaller dots, the macroscopic discoid spots tend to be distributed wreathlike in a circular manner near the pupillary margin or in the zone of the dilated pupil. This appearance was first described by von Graefe. They also are more likely to be found over radial water-slits or dilated sutures. Although it is not possible to see any of the smaller dots in the epithelium biomicroscopically when they are visualized by specular reflection, their closely sub-capsular location does not preclude the epithelial location which has been so well demonstrated histologically. The larger discoid spots are probably formed by a conglomeration of smaller dots, especially in areas where as has also been demonstrated histologically, exuberant growth of the epithelium occurs. The exuberant and stained epithelium may by proliferation extend deeply for a considerable distance in the cortex. This may account for the deeper subcapsular location of the deposits seen with the narrow beam. According to Meesmann the backward displacement of the anterior line of disjunction which he saw in siderosis lentis may also be due to this factor. Macroscopically or with low power it is often seen that zones of discontinuity and the intermediary areas are more blurred than is normal and that their color is more yellowish. This may be due to a "veiling" effect from the subcapsular discoloration. The deeper and posterior parts of the lens are not involved in the process of lens siderosis and hence the ordinary increase of yellowness commonly seen in senility, particularly with nuclear cataract will not be confused with it. Even in cataracta brunescens and nigra, which represent the highest degree of this posterior discoloration in nuclear cataract, the subcapsular zone of the anterior cortex never becomes tinted.

Besides the well-known yellowish discoloration of the iris, it is

interesting to note that both Vogt and Koeppe have stressed the hitherto little mentioned corneal discoloration and vitreous changes (Plate LXXVI, fig. 3). Uthoff (1903) described a similar corneal discoloration using a loupe. Siderosis of the cornea was supposed to be a rarity. This was probably due to the fact that it is difficult to see it by diffuse light. When present, it will be best noticed nearer to the limbus. By comparing the color of the corneal parallelepiped to that of the opposite or to that of a normal eye it will be seen that it has a decided yellowish tinge. The normal corneal block or section is bluish gray in color. The cause of the siderotic discoloration in the cornea is probably to be found in the presence of microscopically yellow to green colored dots within the corpuscles of the corneal stroma. In one case Vogt noted a siderotic line in the corneal epithelium. This resembled the so-called senile corneal pigment line. (See Vol. I, page 381.)

Liquefaction and destruction of the vitreous structure when it is not obscured by lens opacification, may also be a feature of siderosis bulbi. Vogt first observed the presence of vivid red pigment deposits and tablet-like white deposits on the scaffold of the disturbed vitreous. Also, using a red-free light source he found in one case a superficial mosslike area in the fovea from which radiating lines extended (retinal folds?). In addition, with the same light source, in this case of siderosis, there was a decided yellow discoloration of the fundus. He was not certain whether this yellow color was due to the discoloration in the lens and vitreous or to an actual siderotic change in the retina.

Up to the time of the recent work of Itoi⁴⁸⁶ * it was supposed

* Bellows³⁶⁴ citing Itoi's work stated: "The best explanation is that given by Itoi, who was the first to conceive the idea that electrochemical processes are important in the development of siderosis. To test this hypothesis he showed that a nail placed into the eye of a rabbit quickly becomes rusty, while one connected to zinc placed subcutaneously produces no rust. Itoi justly assumes that electrochemical processes account for this phenomenon, since metal can react with its surroundings only when ions become available because of the electrolytic dissolution tension of the surrounding medium. When the iron is connected to the zinc, the flow of the ions is reversed, and no reaction of the surrounding medium is possible. Mielke extended Itoi's experiments in order to explain the accumulation of metal in distant places, e. g., subcapsularly. Not only does he agree with Kohlrausch (1932) that there is actually a constant electric current present in the eye (Bestandstrom), but he believes that it flows in just the right manner to direct deposits to such locations. These deposits may occur at any border surface, such as Descemet's membrane or the lens capsule."

that the epithelium had a special affinity for the oxides of iron. In these cells iron compounds formed which may be nature's way of making the foreign inclusions less toxic.

COPPER IMPREGNATION OF THE LENS

The retention of particles of pure copper within the eye usually causes a severe toxic endophthalmitis, eventually leading to complete ocular destruction. Particles of copper alloys or compounds poor in copper may be tolerated in the eye for long periods but sooner or later tend to produce plating (chalcosis) of the tissues — the cornea, lens, and vitreous being particularly affected. In most of the reported cases in the literature the accident to the eye was caused by explosions, especially of cartridges, grenades, etc., which have containers made up of brass or other copper-containing compounds. The typical picture of chalcosis lentis (sunflower cataract) becomes evident particularly in eyes in which the lens itself suffered no direct injury from the penetrating copper-bearing particle. Although there were a few individual reports of cases having a strange appearance in the anterior part of the lens (Goldzieher [1895],⁴⁵⁵ Ertl [1907]⁴²²) it was Purtscher (1918)⁵⁶⁶ who first established that the so-called sunflower cataract was caused by the intra-ocular presence of copper or substances containing it (Plate LXXVI, fig. 4). Jess (1922-1929)^{488, 489, 490} collected all the reported cases up to 1919 and studied the condition histologically. Vogt (1930-1931) described several cases and gave minute biomicroscopic descriptions of the changes. He listed the following signs as typical:

- (1) Shape. The opacity is ring- or disklike, corresponding in size to a medium pupillary diameter. Opaque peripherally pointed spokes of the opacity radiate from the ring or disk so that a kind of "sunflower" figure is formed.
- (2) Color. Usually the color is grayish green. Hillemanns,⁴⁷³ found it to be of an olive to brownish color; Esser,⁴²³ a fluorescent green; Haab,⁴⁶⁷ gray blue; Bleisch,³⁶⁸ Uhthoff⁶⁴⁴ and Rumbaur,⁵⁸⁶ a brownish or reddish.

- (3) Visibility by retro-illumination. Because of the lack of distinctness when viewed by retro-illumination the opacity was termed an "apparent" cataract.
- (4) Localization. Uniformly found in the anterior parts of the lens (subcapsular).
- (5) Color display. Oblique focal illumination reveals an iridescent display in the shagreen fields of the anterior capsule independent of the underlying gray-green "sunflower cataract" behind it. Vogt believed that this finding is inconstant and is to be seen only in progressive cases.

Owing to the peculiar configuration and color, copper cataract has been likened to and called "sunflower cataract." Some of the earlier writers (Hillemanns and zur Nedden), not having the advantages of the exact method of localization afforded by biomicroscopy, incorrectly localized the position of copper cataract as either on the surface of the lens or in the middle layers of the lens. However, all recent writers agree that its location is subcapsular or in the anterior lens layers (Fig. 448 A, B). The only question still to be decided is whether the opacity involves the epithelium of the anterior capsule or the anterior fibers of the cortex or both. Jess considered the deposit to be subcapsular (cortex) stating that the epithelium resists impregnation of copper and that consequently the salt (carbonate) is laid down in a layer just below it. Vogt stated that with the narrow beam the deposit is everywhere of the same sagittal dimension (being so thin as to defy measurement) and that it is located at the posterior surface of the capsular band or in its immediate neighborhood. Also, there is a clear zone between the opacity and the intact anterior line of disjunction. The opacity did not exhibit any fiber or suture design (see Anterior Cataracta Complicata) which might be expected if these structures were involved.* With low power, the figure is plainly visible as a grayish white and slightly

* In one case of chalcosis, at the time of extraction of the lens Vogt removed a fragment of the anterior capsule and subjected it to the action of hydrogen sulfide. He found black dots (copper sulphide) grouped around the nuclei in the protoplasm of the epithelial cells. The hyaline capsule and superficial lens fibers did not show any black precipitate. Localization of the copper stain with the biomicroscope supports this finding.

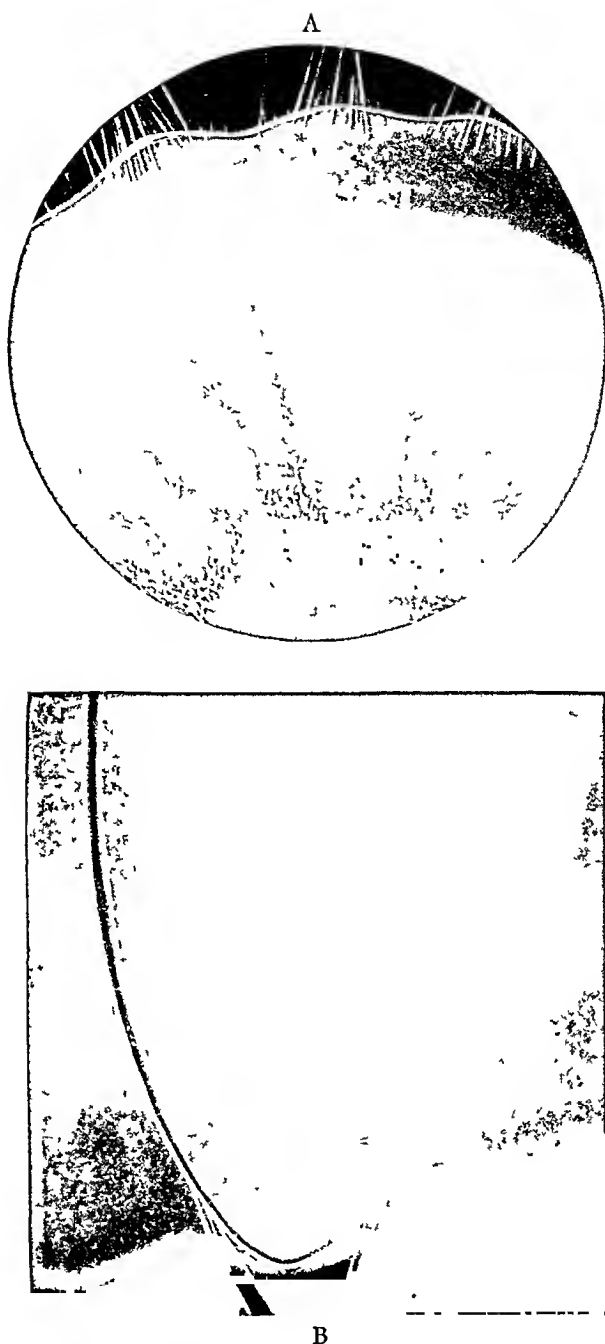


FIG. 448. Sunflower cataract. A. Radiating opacities were slightly greenish yellow in color. Lens dislocated. Edge of lens crenated with zonular fibers missing in retracted areas. Nucleus slightly opaque. B. Optic section showing location of opacities (subcapsular).

tinted (blue, green, or even red) star, its center forming a ring or disk. When a ring is present, the diameter of its clear lumen may measure from 1 to 3 mm. Extending from the central ring or disk are the characteristic rays which taper (retro-iridally) toward the periphery. In Vogt's cases the width of the ray in its middle portion was from 0.8 to 1.12 mm. With high power it may be seen that the entire visible lens area is covered by fine dots but in the region of the figure they are so numerous, condensed and reflecting that it may not be possible to resolve them as such.* In addition to the fine dot-like deposit, Vogt for the first time reported the presence of small dark spots measuring from 0.02 to 0.1 mm. These dark "holes" were seen in the pupillary area in the rosette areas. With varying angulations of the beam they appeared to have a gloss characteristic of flattened vacuoles. No pigmentation of copper cataract has been seen posteriorly. However, as has been noted in siderosis, if backward proliferation of the capsular epithelium at the equator occurred, posterior deposit of copper might follow.

* The exact explanation for the "sunflower" shape of copper cataract has not as yet been determined. A mild controversy concerning this point was raised by Vogt who disagreed with the explanation offered by Jess. Jess supposed that the ring and its processes was brought about by the fact that the parts of the iris which are located on the lens protects the latter from impregnation by copper. Vogt considered the opposite to be true. According to him, the sunflower figure appears just where the iris could contact the lens. He argues that if Jess is correct, then a figure opposite (negative) to the sunflower cataract would occur, i. e., there would be an intensely opaque disk corresponding to the pupil which should be separated from the peripheral radial sectors by a lighter or clear ring. This would be so because of the protecting seam of the iris which contacts the lens capsule at this point. Radial sector opacities would have their base at the equator, and Vogt points out that in reality the opposite is true. The shape of the copper cataract is generally in the form of a ring, which is located where the pupillary border of the iris contacts the lens. The radial processes would correspond to the normal radial (ridges) marking on the posterior surface of iris. These radial folds with their ridges are finer and closer together in the sphincter zone than peripherally where they become coarser; also in the case of the latter their distance from the lens increases (as noted by the greater depth of the posterior chamber peripherally). Consequently the contact of the posterior iris ridges must axially be greater and more extensive than peripherally. It is conceivable that the shape of the many-ridged figure of copper cataract (sunflower), which corresponds morphologically to the relief of the posterior pigmented surface of the iris, is caused by the latter. In a case of copper cataract in a boy aged 11, Vogt counted up to 16 rays in one quadrant. On inspection of a corresponding sector of an excised piece of iris from a normal eye he counted about the same number of ridges. This would also tend to support his hypothesis. From the above it would seem that the iris rather than the aqueous furnishes the toxins that induce this form of cataract. He cites a similar observation of Lundberg who found that in naphthalene cataract opacification did not occur in the region of an iris coloboma. Also confirming this idea, viz., that the development of the shape of copper cataract depends on the iris, is a case seen by Jess in which after prolonged mydriasis, a second ring appeared.

The marked coloration of the anterior shagreen (mirror zone) in which a vivid display of colors, red, blue and green, appears, is not found in every case of sunflower cataract. When present the color display, not unlike that seen in complicated cataract, will be found even in areas outside those occupied by the underlying sunflower figure. Hence it must be considered as a phenomenon independent of the cataract itself.

In addition to discoloration of the iris, vitreous changes are seen. The vitreous may be more fluid than normal. This is indicated by the presence of large clear (dark) spaces and by the increased motility of vitreous trabeculae upon movements of the eyeball. In such cases the vitreous may exhibit a gray-green haze. This haze is due to the presence of dots and dust of great delicacy. These dots are smaller than those seen after inflammation (leukocytes). At times the gray-green discoloration of the vitreous, which usually gets more pronounced in its deeper portions, even surpasses that of the lens.

SUNFLOWER CATARACT IN HEPATOLENTICULAR DEGENERATION^{*} (WILSON'S DISEASE)

In association with Kayser-Fleischer ring (Vol. I, page 385) a typical sunflower cataract may be found in cases of Wilson's disease. Vogt considered that the corneal and lenticular pigmentation in this disease may be due to a copper or silver intoxication.* This followed in line with the previous conception of the causation of the Kayser-Fleischer ring of Wilson's disease as advocated by Rumpel, Spatz, and Siemering. According to Duke-Elder, their theory was "that in hepatolenticular degeneration the body failed

* Vogt stated that the solubility of the pigment in potassium cyanide indicated its metallic nature. Duke-Elder points out that solubility in potassium cyanide is a property common to other metals and to organic pigments as well. Adherents to the idea that the deposit is organic rather than metallic (Jess, Gerlach and Rohrschneider) brought some evidence in favor of this theory. Jess showed that the pigment fades after long embedding in celloidin (a feature common to organic pigments) and the others were not convinced after spectrographic examinations that it was metal. In a recent biomicroscopic survey, I found that during the stage of icterus and for some time afterward it was possible to detect a faint but definite discoloration (greenish blue to reddish) at the level of Descemet's membrane in the periphery of the cornea. It never was as marked as that seen in Kayser-Fleischer ring but at the same time this finding is suggestive. In none of these cases was there any evidence of a sunflower cataract.

to dispose of the metals ingested in food, and that these (copper and silver) were deposited on Descemet's membrane, while the corpus striatum had a peculiar chemical affinity for them." Originally both Fleischer and Hall considered that the pigmentation in the cornea was hematogenous. Poe (following Kirbik, Hessburg, and Jess) believed it represented derivatives of bile pigments. As in the case of copper cataract, the sunflower cataract in Wilson's disease may be missed unless searched for carefully with the biomicroscope. Vogt originally found this condition by chance in a case that was being refracted. The opacity has the same morphologic aspects, localization, and color as does copper cataract.

MERCURY IN THE LENS (HYDRARGYOSIS LENS)

Recently Atkinson,* reported for the first time the finding of a colored reflex from the anterior parts of the lens in cases of chronic mercurialism. The importance of this finding from the standpoint of industrial medicine as a diagnostic feature of mercurialism cannot be overemphasized. He stated that "the color of the reflex is deeper in the pupillary area, and in different individuals its color varies from a light brownish gray to a deep rose-brown. The depth of color apparently depends somewhat upon the length of time and the amount of mercury to which the individual has been exposed. Small, round, punched-out defects and cracks often appear in the reflex. These resemble the behavior of metallic mercury. Glass lenses that have been exposed to mercury often present a similar brown reflex when examined with the slit lamp." The posterior cortex and capsule were unaffected. The presence of reflex seemed to be a constant finding (see Fig. 448 C). The deposit does not interfere with vision. It is not apparent ophthalmoscopically and depending on its intensity appears as a delicate dull gray haze when examined by ordinary oblique illumination. Intention tremor, affections of the teeth and gums and skin pallor were seen in varying degrees. In a later communication,† Mercury in the Lens, histological, histo-

* Atkinson, W. J., *Tr. Am. Ophth. Soc.* 40:254, 1942; *Am. J. Ophth.* 26:685, July 1943.

† Atkinson, W., and Sallman, L. von, *Hydrargyrosis Lenti*, *Tr. Amer. Ophth. Soc.*, 1946.

chemical, and spectrographical studies were made of lenses removed from a patient with chronic mercurialism in which the colored reflex was present. Although biomicroscopically the staining appeared

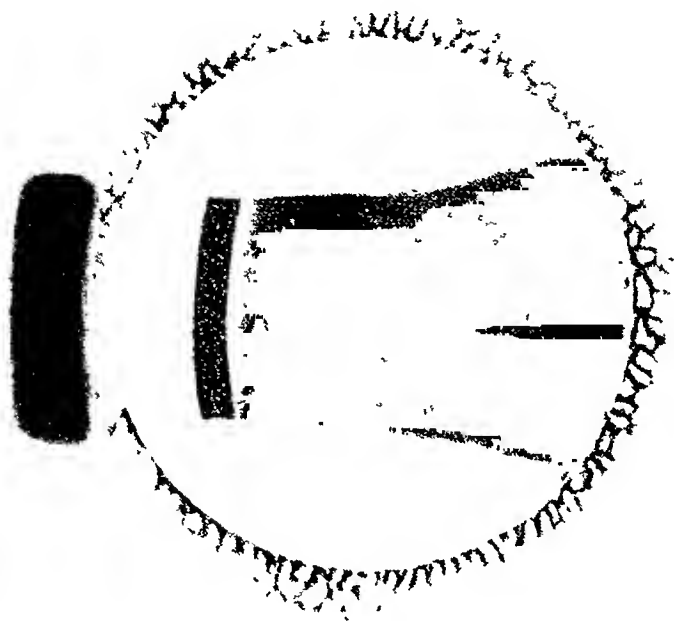


FIG. 448 (Continued). c. Cataract in mercury poisoning.

capsularly or just subcapsularly, in the prepared sections the lens capsule and epithelium did not contain the deposits and were in general normal. Sections of the lens fixed with Cristeller solution showed black or dark brown particles located in the superficial cortex, more numerous in the equatorial regions despite the fact that with the biomicroscope during life the brownish reflex appeared more dense in the pupillary area. The mercurial nature of these deposits was also confirmed by additional microchemical tests. Control lenses similarly fixed were free of such deposits. Spectrographic examination of one of the lenses showing the reflex revealed traces of mercury whereas no mercury was detected in control lenses.

Chapter Twenty-Eight

RADIATION CATARACT

IN 1800, Herschel showed that besides the visible part of the spectrum there existed an invisible or "dark" part. The visual part of spectrum, that part which excites the sensation of light in the retina, occupies only a small band in the total spectrum. The invisible part of the spectrum is also subject to the laws of reflection, polarization, and refraction. The wave length of the visible spectrum (red to violet) ranges between 7,000 to 3,500 angstrom units. Above this are the infra-red while below are the ultraviolet, roentgen and gamma rays, etc. Fundamentally as far as the tissue of the eye is concerned only those rays which are absorbed are harmful to it (Draper's law, 1879). Those that are transmitted or reflected excite no reaction.* The passage of even intense visible light (free of dark radiant energy) through a plate of glass or water will not raise the temperature of either since no light is absorbed. The same holds true for the eye media. But if a piece of black paper is put behind the above mentioned plate of glass the temperature of the glass will rise owing to the absorption of the light by the black

* Duke-Elder succinctly explains the affects of absorbed radiant energy as follows: "The periodicity of the longer wave lengths—the infra-red—may be conceived as corresponding in a general way with the movements of the atoms. They are thus absorbed into this periodic system, and their energy finds expression in an increased rate of atomic movement, a phenomenon which is appreciated as heat, and which ultimately results in coagulation of the proteins of the tissue, thus causing a thermal lesion. The radiations of intermediate wave length—the visible rays—finding no sympathetic resonation, as it were, slip through the transparent media without exerting any affect until the retina is revealed. The shorter ones—the ultraviolet—whose frequencies correspond to the intra-atomic periodic oscillations, are again absorbed. The energy they represent is added to the atomic system so that the electrons are made to change their orbit, thus altering the chemical nature of the atom, or are jerked out of the atomic system altogether, thus producing a photo-electrical effect; this results ultimately in coagulation of the proteins and the production of an abiotic lesion. Both these—the thermal and the chemical—are quite distinct, although in the last analysis they are identical, and are dependent upon the transference of energy to the molecules or their parts, with the result that they are shaken apart."

paper. By contact the glass gets heated. This may be compared to the process that may occur in the pigment epithelium under the retina when the eye is exposed to intense visible light.

Fortunately, owing to the absorptive power of the cornea and lens, only a small part of the invisible rays ordinarily present in our milieu (ultraviolet and infra-red) is able to reach the retina. However, quantitatively this small part (especially the short infra-red rays) actually exceeds in amount the visible rays. The cornea absorbs all radiation above 20,000 Å. U. (long infra-red or heat rays) and below 2,900 Å. U. (short ultraviolet).^{*} Consequently when of sufficient intensity, the former can induce a thermal lesion and the latter an abiotic photochemical lesion. The degree of abiotic reaction varies directly with the time of exposure and inversely as the square of the distance of the source of light from the eye.

That exposure to solar light under certain conditions can result in transient or permanent ocular damage has been known for a long time, e.g., eclipse and snow blindness. Vogt considers that both visible light and short infra-red rays play a role in the causation of eclipse blindness. In pigmented tissues all incident energy is absorbed indiscriminately and is degraded into heat (Duke-Elder). Hence both visible light rays and the short infra-red rays could take part in heating the underlying pigmented epithelium. This in turn by radiation could act thermically on the overlying retina. Vogt experimentally was able to produce retinal "burns" by means of short infra-red rays alone. Since only an insignificant amount of ultraviolet reaches the retina, it is highly probable that this form of energy does not cause the macula alterations that follow gazing at the sun or at an eclipse. Snow blindness or burn (affecting the cornea and conjunctiva—especially the epithelium) on the other hand is directly due to the effects of the shorter ultraviolet radia-

^{*} The cornea transmits most of the heat rays (infra-red) ranging between 20,000 and 7,200 Å. U. and hence these rays can reach the lens through the pupil. The lens only absorbs a small percentage (12 per cent) of these rays—particularly those between 13,500 and 11,000 Å. U. When these are intense a heat cataract can develop. In addition, the cornea transmits roentgen rays and gamma rays. These rays have extremely short wave lengths—hardly more than 1 Å. U.—and under suitable conditions may induce lens changes. Cataract from this form of radiation appears only after a long latent period.

tions. At sea level only the longer ultraviolet rays predominate, i.e., from 3,500 to 3,100 Å. U. This radiation in ordinary and normal amounts does not cause any damage, because ontogenetically and phylogenetically it has become "physiologic" for our eyes. However at higher altitudes — especially above 6,000 ft. — ultraviolet radiations much richer in rays of shorter wave length (2,900 Å. U.) are found. It is these which cause burns of the conjunctiva and cornea. Likewise the abiotic affects to the cornea and conjunctiva after exposure to artificial light (ultraviolet emanations from mercury vapor and arc lamps) is well recognized.

The lens is especially vulnerable to short-wave heat rays, absorbing a considerable amount of the infra-red between 14,000 and 11,000 Å. U. This is important because 75 per cent of the rays in this band striking the cornea pass through it (Hartridge).

The cornea also transmits the longer ultraviolet rays between 4,000 and 3,200 Å. U. and although, experimentally, histologic evidence has been brought forward to indicate that in so doing damage to the capsular epithelium of the lens may result (Verhoeff and Bell, 1916; Duke-Elder, 1929, and others), the consensus today holds that, practically, ultraviolet energy is not deleterious to the lens. Because of the fact that the normal lens appears whitish in ultraviolet light, earlier investigators thought that this might play a role in the production of senile cataract. Clinically well-defined cataracts have not been produced even after prolonged exposure to ultraviolet rays. Actually it has been pointed out that in persons living in higher altitudes (richer in ultraviolet) the incidence of senile cataract is not greater than in those living at sea level. In addition senile cataract starts in the peripheral parts of the lens which is protected from the direct affects of radiation by the iris.

Fluorescence. In addition to the properties exhibited by radiant energy already discussed (transmission, absorption, reflection, etc.) the phenomenon of fluorescence remains to be considered. In the eye, this phenomenon is associated especially with the lens. Simply, it means that the lens has the power of changing the wave length of part of the radiant energy incident upon it. Fluorescence in the lens

occurs when it is subject to the long ultraviolet rays whose length is from 4,000 to 3,000 Å. U. — the greatest effectiveness being from 3,900 to 3,700 Å. U. When the lens is irradiated with light of this wave length (in a darkened room) it gives off a peculiar greenish-yellow glow.

Duke-Elder⁴⁰⁰ states: "Here (in the lens) part of the light gives rise to *fluorescence*, the region of greatest effectiveness being in the long ultraviolet, from 3,500 to 4,000 Å.U. This is a phenomenon whose intricate nature is little understood. When light passes through a fluorescing substance, the particles of the substance become light sources themselves, emitting light of a wave length differing from that of the incident light, preponderatingly longer but some shorter. The effect of this phenomenon is regarded differently. Considering that its production implies absorption, Schanz (1915-22) looked upon it as deleterious. On the other hand, from the work of Bunge (1915) who correlated a decrease of fluorescence with an increase in coagulability of lens proteins, it may possibly be looked upon as a protective mechanism, whereby active short waves, which might induce coagulation, are changed largely into long (visible) waves, by which means their energy, being in a more transmissible form is disposed of more safely."

Vogt (1913)⁶⁴⁵ showed that this phenomenon could also occur to a lesser degree in violet and blue light (visible light) as well. He noticed that in violet light the hue of the fluorescence varies with the changing color of the normal lens with age. In the young, in whom the lens is almost colorless, the fluorescence is bluish white.* In the case of old people whose lenses become yellowish, especially in the posterior cortex, the fluorescence — appears yellow-green in this light. The yellow color acts as a filter which screens out the blue and violet (cold) hues and passes the warmer ones.

In ultraviolet light the aged lens does not fluoresce as effectively as the young one. In the former the color is faintly yellow, while in the latter it is definitely greenish. The physiologic increase in yellowness with age tends to offset the loss of the power of fluorescence; in this way protection against the undue action of active short waves is effected.†

* Even in young humans the lens is not entirely colorless and transparent. Calves' lenses, on the other hand, being highly transparent, do not fluoresce in violet light.

† In this connection, Wald has shown that aphakics have a 1000 per cent increase of visual acuity in ultraviolet light as shown by their ability to read an ultraviolet illuminated Snellen chart invisible to a normal observer (phakic).

HEAT ("FIRE") CATARACT; INFRA-RED CATARACT

That cataractous change can occur from exposure to the relatively short-wave infra-red, penetrating radiation from a glowing mass of glass or metal is now a well-established fact (Glass Blower's and Foundry Worker's Cataract). Vogt, who did considerable experimental work in the field (1911-1921), showed that the damage to the lens (in experimental animals and man) resulted not from the ultraviolet rays, visible light or the very long heat rays, but from the abundant and predominating infra-red rays (7,500 to 3,400 \AA . U.), which emanated from these heated and glowing masses.* Hence it is not surprising that men engaged in occupations that expose them to this kind of irradiation (glass blowers, foundry workers, blacksmiths, welders, etc.) are liable to suffer from lens changes. As will be seen, these changes form a specific type of cataract, and hence they bear the name "glass blower's" or "fire" cataract.

The amount of ultraviolet and visible light that emanates from these sources is less than that found in ordinary daylight and hence can be considered of little importance. In 1918 Vogt succeeded for the first time in making an apparatus with special filters and optical system that made it possible to obtain short-waved infra-red rays in sufficient quantity. After an exposure of three hours, cataract developed in experimental animals (rabbits). With strong doses he was able to produce anterior lens changes as well as the characteristic

* Two kinds of invisible infra-red (heat) irradiations must be recognized. First, the long (nonpenetrating) waves, those over 24,000 \AA . U. and second, the shorter waves, from 24,000 to 7,500 \AA . U. The long-waved rays are absorbed by the tear film and the corneal surface. Exposure to them results in a sensation of warmth and eventually if intense to conjunctivitis and keratitis (thermal reaction). Long nonpenetrating infra-red is radiated in large amounts by stoves and sources of artificial light (incandescent bulbs). The shorter infra-red rays predominate in glowing masses of glass or metal. Vogt showed that these pass through the cornea and are absorbed by the iris and especially the lens. Hence he called them "penetrating infra-red rays." In other words it is only a heated body giving off a red glow (500 degrees R or over) that is able to radiate infra-red rays sufficiently small in wave length to penetrate the media and affect the lens. Practically, the fact that persons working in temperatures even higher than those in which glass blowers or foundry workers are subjected to, do not develop the typical "fire" cataract speaks for the specificity of the shorter "penetrating" infra-red rays.

posterior cortical changes. Cataract developed more readily in the older animals and in those that were pigmented. The former findings coincide with the fact that most of glass blower's cataracts in humans were found in older persons; not entirely because of greater number of years of exposure but rather that old lenses absorb more of the infra-red rays. This is further evidenced by the frequent finding of nuclear changes (cataract nigra and brunescence). The average age for the appearance of glass blower's cataract varies between 55 and 60 years. This is considerably lower than that found in senile exfoliation of the capsule. Because it was easier to produce cataract in pigmented animals (although later Vogt did so in albino animals also), the famous controversy between him and Goldmann arose.* Vogt considered that the action of the short infra-red rays was specific to the lens. Goldmann⁴⁵² held that the lens changes were secondary to overheating of the iris and also to increase of the temperature of the posterior chamber following the rise of general body temperature. He based his contentions on the difficulty of producing cataracts with exposure to infra-red in albino rabbits and even in the pigmented species when the iris was protected. Vogt admitted that the heat absorbing power of the pigmented iris could play a role in raising the temperature in the anterior chamber and consequently that of the cornea, but felt that this would only augment the direct action of the irradiation to the lens. Experimentally and clinically the point that the lens changes in the beginning are located in the pupillary zone both anteriorly and posteriorly, and that the neighboring ocular tissues (cornea, iris, and vitreous) are manifestly not affected, also tends to confirm Vogt's

* The question has been raised whether the type of changes produced in animals by large single or intensive doses are comparable to those found in glass blowers and foundry workers who are exposed to fractional doses, extending over long periods. (In practically all the reported cases the characteristic lens changes of glass blowers and foundry workers cataract never appeared in individuals before 15 or 20 years of employment in these occupations.) Clinically Vogt was able to produce by irradiation (in a case of an intra-ocular tumor) with short infra-red rays the typical changes of heat cataract. The eye was irradiated with fractional doses for long periods from one direction—so that the one and the same part of the lens was exposed to its effects. After the fourth treatment typical anterior subcapsular homogeneous dust-like opacities appeared in the area irradiated. With further exposure the opacities thickened and six months later the whole opacity was pushed deeper into the cortex by new clear fibers. Meesmann, employing small and repeated doses of infrared in rabbits produced axial cataract in the posterior cortex—similar to that found in glass blower's cataract.

idea that the damage results principally from the direct action of the rays on the lens. From the standpoint of biomicroscopy the alterations in glass blower's and foundry worker's cataract may be enumerated as follows:

- (1) Detachment of the superficial lamella of the anterior lens capsule (*solutio laminae superficialis*)
- (2) Posterior and anterior axial subcapsular opacities similar to saucer cataract
- (3) Superficial (anterior) gray radial striae (wreathlike opacities [Schläfen])
- (4) Nuclear cataract (especially *cataracta brunescens*)

DETACHMENT OF THE SUPERFICIAL LAMELLA OF THE ANTERIOR CAPSULE

Although detachment of the superficial layer (lamella) of the anterior capsule (*Solutio Laminae Superficialis* — Fire Lamella) is not found in every case of heat cataract, when present it is diagnostic of glass blower's cataract. It has not thus far been reported in any other condition. And, like senile exfoliation, it has never been found on the posterior capsule. Occasionally it may be found long before any other lens changes are noted (especially the characteristic anterior or posterior polar opacities), but in most instances it is accompanied by cataractous changes of varying degree.

Of the cases reported in the literature, the ages of the patients in whom fire lamellae were found ranged on the average between 50 and 60 years.* The youngest (Schnyder⁶⁰³) instance occurred in a 42-year-old smelter.† Elschnig⁴¹⁸ was the first to describe the detachment and to interpret it correctly as an occupational disorder in glass blowers. However, he called it detachment of the zonular lamella; today it is realized that it probably does not involve the zonular lamella, the location and attachment of which is more equa-

* By comparison it will be seen that the age range for senile exfoliation (which though affecting similar parts of the capsule is morphologically different in appearance) is notably higher.

† In the case of smelters or those exposed to the glow of liquid metal the earlier appearance of fire lamella (and other changes) may result from the higher temperature and more intense irradiation of penetrating infra-red rays to which they are exposed.

torial (page 1331). Meesmann first described the appearance of an actual detachment of the zonular lamella after trauma (page 1332). Actually the part of the capsule that separates in heat cataract starts

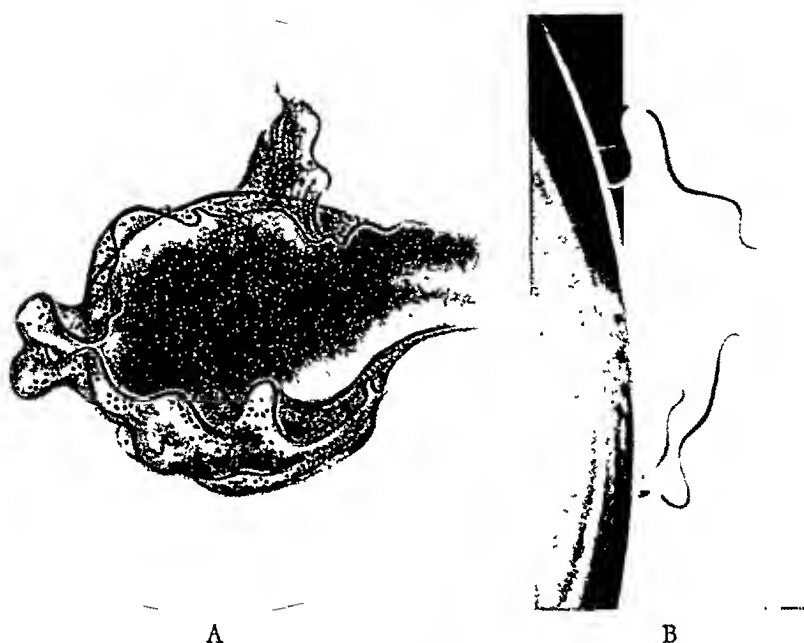


FIG. 449. Heat cataract. A. Frontal view. B. View by optic section.

just at the place where the zonular lamella ends (a weak transitional zone) and extends axially in a more or less concentric way retro-iridally nearly to the pupillary zone (Fig. 449 A, B). Just as in senile exfoliation, a somewhat opaque pupillary disk may exist. As mentioned previously, the morphologic difference between fire-lamella and senile exfoliation may only be one of degree. In senile exfoliation, the separation is more delicate and is characterized by fine crumbling powdery or gray flaky particles. These particles, being extremely light in weight as compared to their size, float freely like inflammatory precipitates in the currents of the aqueous; they may become attached to the pupillary margin and in certain instances may clog up the filtration angle and produce increased intra-ocular pressure (glaucoma capsulare).

In detachment of the superficial lamella in heat cataract the separated pellicle is thicker, continuous, and like a glass membrane it is reflecting. (See detachments of Descemet's membrane, Vol. I, page 422.) One might compare the detached portion, which is directed

toward the periphery,⁶⁰¹ to a transparent sail which can be agitated in the anterior chamber by eyeball movements (Fig. 449 A). The detached membrane may be folded; in a case described by Vogt the dorsal surface was sprinkled with pigment.

In senile exfoliation the ends of the separated pellicle are directed axially. In contrast to senile exfoliation in which the detached portions are opaque and easily visible, it is usually impossible because of its transparency to decide exactly at what point the fire lamella has become detached. Axially the detached membrane may extend up to a pupillary disk, and the edges may or may not be raised. Schläpfer also reported in glass blowers the presence of a fine line running concentric to the dilated pupil (without definite separation of the lamella). This line, which may represent the beginning of rupture or separation of the lamella, was delicately pigmented.

The first histologic preparation of fire lamella was made by Vogt who studied two cases. He found that the separated lamella (which in one case was about one-half the thickness of the entire hyaline capsule) contrary to the usual biomicroscopic appearance, varies in thickness. However, Elschnig in his biomicroscopic description found that lamella is not homogeneous but is composed of fine parallel and horizontally directed lines or subdivisions (parallel stripings). This form of laminated striping or separation of the hyaline capsule into subdivisions has also been seen in histologic preparations of other types of cataract (e.g., senile) and suggests solution of the cement substance between the laminations. Evidently such a process, while it is not specific, is more marked in the fire lamella. The condition of the underlying epithelium varies. In some cases its nuclei are deformed, and the cells are fusiform. The underlying lens fibers exhibit varying degrees of vacuolar destruction.

ANTERIOR AND POSTERIOR SUBCAPSULAR SAUCER-LIKE OPACITIES

The fact that subcapsular opacities (especially those of the posterior pole) are significant for heat cataract was proved experimentally by Meesmann and by Vogt. By using fractional doses of penetrating infrared over long periods of time in experimental animals (rabbits), Meesmann was able to produce posterior polar lens

opacities and thus was able to simulate this characteristic type of opacity so frequently found in glass blower's cataract.* Using more intense irradiation over shorter periods of time Vogt also demonstrated experimentally in rabbits that cataractous changes appeared first where the light bundle contacted the lens (i.e., the anterior cortex) and then where the rays leave the lens or the posterior cortex. In his animals, homogeneous fine dustlike opacities appeared in the anterior and posterior polar regions soon after irradiation. Larger vacuoles only appeared much later. The opacities preferred the region of the sutures (a finding seen in many other types of cataract, i.e., complicata, senilis and traumatica). As a rule after an early progression and providing total cataract does not occur the infra-red (experimental) opacities become stationary. However, as in traumatic cataract, the layer of opacities may be pushed deeper into the cortex by the ingrowth of superficial clear fibers. With this type of irradiation the nucleus was relatively immune. In a human subject (vide supra) Vogt was able to produce similar circumscribed and permanent opacities.

The finding of subcapsular (polar or parapolar) opacities as a manifestation of glass blower's cataract has been verified by numerous authors. Ocular complications, i.e., iridic changes, precipitates in the anterior chamber and especially in the vitreous, are absent in heat cataract. This again would tend to strengthen the proposition advanced by Vogt and his school that strictly speaking infra-red cataract is not primarily a complicated cataract in the sense that it is caused secondarily from the effects of heat on the surrounding tissues. The differential diagnosis between senile saucer-shaped opacities (in the absence of anterior lamella separation) may be difficult. Also since most heat cataracts occur in older individuals, the presence of well-developed ordinary senile cortical and nuclear

* The opacity was radial to sutures of the posterior cortex. Vogt believes that this area is especially vulnerable. Legge cited by Bellows points out that "the region of the posterior pole is involved first because the waves are refracted by the cornea and lens and become more concentrated there. This concentration is enhanced by some reflection from the concave posterior surface of the lens." In the experimental animals the opacities were in the beginning dust-like and with higher power seemed to be of vacuolar nature. Similar to traumatic cataract (remotio) and zonular opacities, Vogt found that after some months they were progressively displaced deeper and deeper into the cortex.

cataract may so obscure the picture that, as mentioned above, in the absence of anterior lamella separation a differential diagnosis from the standpoint of morphology may well be impossible. One point may be brought out in this regard — it is not common in senile saucer cataracts in the early stages for subcapsular opacities to be limited to the polar regions.

Both Schnyder and Vogt have emphasized that posterior opacities, in heat cataract may be arranged in a double or triple layer (meniscus form). Each successive layer from before backwards increases in density so that as a rule the posterior one is the thickest. Each layer, composed of vacuoles and flaky material, is thickest axially and flattens out peripherally where, as small dots and lines, it gradually loses itself in the clear lens substance. Frequently, the separation of these layers by lucid intervals is greatest axially, the peripheral ends tending to converge. The more delicate and most anterior of these layers usually does not extend as far peripherally as do the deeper ones and consequently, in diffuse light circles of opacities may appear. Considering the age of these patients it is not surprising that the posterior subcapsular opacities are commonly tinged yellow.

The anterior subcapsular opacities are more delicate and in the beginning are composed of fine dots, which under higher powers of observation and retro-illumination may have a vacuolar appearance. Larger clear vacuoles are seen frequently in association with the finer opacities. As is common in anterior saucer opacity the changes at first appear (under low power) as a small area of axial blur with one or more radial processes or stripes extending from it. These radial stripes apparently follow the direction of the lens fibers. With the narrow beam some of the finer deposits may be directly subcapsular but most often they will be found condensed at the anterior disjunction line.

SUPERFICIAL (ANTERIOR) GRAY RADIAL STRIAE

In 1930, Rehsteiner and Schläpfer⁶⁰¹ described the presence of superficial gray radial striae situated about midway between the lens vertex (axial) and the equator (Fig. 450). Unlike the "fire"

lamella they are not specific for heat cataract. Schläpfer considered the striae as a new finding in glass blower's cataract. These authors found them in slightly more than half of a series of 59 glass blowers



FIG. 450. Heat cataract. Radial striae. (After Vogt.)

and only in a fifth of persons whose occupations did not entail exposure to infra-red. In contrast, Abramowicz reported these striae in 90 per cent of 240 persons, all of whom gave no history of occupational or other prolonged exposure to heat. Dilatation of the pupil is usually necessary to observe the striae. The stripes are somewhat poorly defined and hence can be overlooked easily. When the entire pupillary area is observed by strong diffuse light, it will be seen that they form a ring or wreath more or less concentric to the pupil. The ring is composed of delicate radiating gray lines, i.e., running at right angles to the pupillary margin. These lines especially those which are nonpigmented, are reminiscent of the retro-iridal lines (page 1023).

Vogt localized the striae as subcapsular since they were covered by the shagreen of the anterior capsule. Since the significance of these radiating structures is not entirely understood, we can only consider them as pertinent to heat cataract when a positive history of exposure to the prolonged action of penetrating infra-red is established.

NUCLEAR CATARACT

Most of the observers of heat cataract agree that nuclear sclerosis and nuclear cataract are features of this syndrome. The sclerosis, which may involve the deeper cortex, may explain the subcapsular location of the opacities mentioned above, since it could prevent further extension into the cortex. Also related to the sclerosis and nuclear opacification is the increased yellow discoloration of the deeper parts of the lens. (This causes the yellow or brownish color of the posterior cortical opacities which necessarily are viewed through it.) In heat cataract, the discoloration seems to precede by a considerable amount of time that found in very old persons.* It should be noted that the appearance of the nuclear cataract in this condition differs in no way from that ordinarily seen in senility (page 1147). Nuclear opacification (especially the rubra type) in glass blower's cataract at times may precede the formation of the well-known posterior saucer opacities with which it is usually associated. One of the advantages of the narrow beam as afforded by the biomicroscope is the ability to detect early clouding in the nucleus. It begins as a grayish condensation within the fetal nucleus and gradually extends outward in both directions. Later it is characterized by a more diffuse central haze, which is separated from the adult nucleus band by a clear interval. The tendency toward deep discoloration (posterior cortex) is marked (*cataracta brunescens* and *rubra*) with a gradual transition to yellow and greenish shades anteriorly. However, nuclear cataract by itself can in no wise be considered as a form of heat cataract unless a positive history of exposure to infra-red, other changes such as anterior and posterior polar opacities and, above all, fire lamella are present. Only the last finding is irrefutably diagnostic.

* According to Berner the absorption of infra-red in the nuclei of lenses in old horses and cows is much greater than that of young animals. This could explain the reason for the nuclear involvement in glass blower's cataract since it generally was not found in the experimental infra-red cataracts in growing rabbits. Perhaps the young lens containing a high percentage of water absorbs less infra-red than does the older lens with its sclerosed nucleus.

ROENTGEN (X-RAY) AND RADIUM CATARACTS *

The most distinguishing features of the lens changes following irradiation by x-rays or radium lies in a prolonged latent period (one-half to one year in experimental animals and from 9½ months to 8 years in man) between the time of exposure, the appearance of cataract, and the progressive nature of the opacities themselves. These two factors have been verified both experimentally and clinically. In this way radiation cataract differs markedly from infra-red cataract. In animals, lens opacities preceded by ephemeral iritic irritation appear a few days after infra-red irradiation. Whereas the opacities (very similar morphologically to those of infra-red) following x-ray and radium first become visible only after many months, even in cases in which the exposure has been sufficient later to produce total cataract.

The failure of earlier authors to recognize the vulnerability of the lens to this kind of irradiation lay in the fact that they were unaware of the long latent period. The older observers were hampered by the lack of the biomicroscope. With this instrument it has been possible not only to observe the finest alteration but also to determine whether they are progressive or not. Vogt has stressed the important differential diagnostic point between heat and x-ray cataract. In the case of heat cataract, provided a total cataract does not form immediately, the opacities do not tend to progress but rather may be displaced deeper by new anteriorly placed clear fibers (similar to remotio cataract). Whereas x-ray and radium cataracts, when once started, continue to progress and advance deeper — an indication of epithelial devitalization.

It has in addition been demonstrated experimentally that relatively, the young lens is more sensitive to the action of these rays

* Since the effects of both these forms of radiant energy on the lens are similar, they will be considered together. In eight experimental trials (Schiez cited by L. Peter⁵⁶¹) there were no morphologic differences found between the cataracts caused by radium or x-rays. Bellows, citing Desjardin, states: "Histologic alterations likewise are quite similar except that degenerative changes in the blood vessels are more prominent after radium because the slight distance between the radium and the eye permits the beta rays to exert their intense short-range influence."

than the lens in older animals. This seems to be directly opposite to the effect seen after infrared irradiation where absorption in the lens increases with age. Whether the damage to the lens from x-rays and radium is direct or secondary through their effect on the ciliary body is still unknown. Because the opacities appear axially in the posterior subcapsular region some have leaned toward the latter conception. This would not explain the frequent early finding of anterior subcapsular changes. Histologically Vogt was unable to find any changes in the equatorial regions. Reese in a histologic study of 16 radiation cataracts removed intracapsularly in humans, found migration of the epithelium posteriorly at the equator. He confirmed the findings of others, i.e., that zones of subcapsular (cortical) vacuolization and fiber destruction occur. In addition he found dense plaques of subcapsular scars resulting from metaplasia of the epithelial cells.

Judging from the illustrations in the literature of experimental x-ray and radium cataracts there appears to be little difference in the morphologic aspects between them and the changes in animal lenses resulting from exposure to penetrating infrared (or some of the other exogenously caused cataracts). In both, the opacities are fine and are located subcapsularly predominantly posteriorly. However, Vogt has pointed out that in infrared cataract the opacities are more dustlike and homogeneous.

BIOMICROSCOPIC APPEARANCE OF X-RAY AND RADIUM CATARACT

According to the cases reported in the literature there seems to be considerable variation not only in the morphologic aspects of the lens lesions but also with regard to the dosage of radiation necessary to produce them.* Although the tendency to cataract seems less in

* Lina Peters, who reviewed the experiments of Sching and Rados (who were unable to find opacities—possibly because they did not observe their animals for a sufficient length of time nor did they present biomicroscopic findings) was able to produce cataracts in rabbits (6 months to 1 year old). She employed doses varying from 1 to 10 skin erythema doses (1.5 S.E.D.=550 r). As is the case in heat irradiation, an ephemeral exudation reaction appeared in the anterior chamber within three days but cleared promptly. It was not until several months later (this interval depending on the strength of the irradiation) that typical posterior and anterior subcapsular opacities and vacuoles appeared.

Numerous reports are at hand concerning the dosage and latent period of irradiation by

adult lenses this does not mean that they are immune. Depending on the length of irradiation, changes appear even in the aged lens. Whether or not irradiation hastens the progression of minute and

incipient senile cataractous changes is as yet not known. According to Milner, the gamma rays of radium are more damaging to the lens than are the beta (undoubtedly because of the lesser penetrating power of the latter, since the beta rays are actually far more destructive to tissue).

With the biomicroscope the early lens opacities, like those in heat cataract, appear in the axial region subcapsularly and predominantly posterior. However, no strict rule can be made because in some instances changes are first observed in the anterior polar regions. The most characteristic lesion is that of a thick dense disklike opacity located subcapsularly or deeply cortical at the



FIG. 451. X-ray cataract.

posterior pole (Fig. 451). This disk, seen best in its entirety by strong diffuse illumination, may be composed of one or more layers (one in front of the other) so that from in front they appear like concentric rings. With the optic section this layered structure when present is seen easily. The opacity is usually densest in the center, gradually fading out in the form of small white points (sometimes as radiating processes) in the clear surroundings. Peripherally the edges may converge toward one another and fuse, tending to take the shape of a flattened convex lens. As already mentioned this meniscus form of opacity is not specific for x-ray or radium cataract but may occur in

x-ray and radium in which cataract appeared in man. The minimal dosage that is able to produce cataract in man is still unknown. The differences in this regard being so great suggest individual variations and susceptibilities. Clapp collected 34 cases in the literature, the latent period of which varied from 9½ months to 8 years. Vogt cited three of his cases aged 7, 40 and a child of 12 years, all of whom were irradiated because of tumor or suspicion of tumor. The doses were 4.5 S.E.D. applied in three to four sessions. In all, posterior subcapsular cortical opacities, followed later by anterior ones, appeared in at least one year following the last irradiation.

posterior saucer (senile) cataract, in complicated cataract and in glass blower's cataract as well. In composition, the opacity resembles saucer-shaped (senile) cataract rather than *cataracta complicata*—being composed of a mixture of fine crumblike opacities and vacuolar structures. Like saucer-shaped cataract and unlike complicated cataract, the opacities tend to be limited to definite layers, the borders of which (anteriorly and equatorially) are sharply demarcated. In older individuals, as is to be expected, these posterior cortical opacities usually have a yellowish tinge. Anteriorly the axial sub-capsular opacities vary from case to case. Usually, they closely resemble those described under the heading of anterior saucer-shaped cataract (page 1141). Being composed of delicate dots which by retro-illumination have a vacuolar appearance, they have a definite tendency to radiate (according to the sutures and direction of the fibers). In extreme cases condensation of the anterior opacities in the middle layer of the cortex may also result in the formation of a dense central anterior disk (Vogt, Peters, Meesmann and Reese). In one case, in a young girl whose eyelids were irradiated for vernal catarrh, the cataract became total two years following a single course of four screened radium treatments. After the second treatment several small skin and conjunctival telangiectases appeared. There also was loss of cilia and a mild eyelid dermatitis. In other cases keratitis, developing into an intractable keratitis bullosa have been noted. All in all one should be extremely cautious in the employment of these modalities (x-ray and radium), especially in cases where simpler and less dangerous therapeutic measures are available.

ELECTRIC CATARACT (CATARACTS FROM LIGHTNING AND ELECTRIC SHOCKS)

Cataract which results from the effects of natural electricity (lightning) differs in no way from those following electric shock with ordinary ranges of generated voltages. Recently a comprehensive discussion of electric cataract was given by Gabrielides (1935) and Skydsgaard (1939). The former reviewed the published cases from 1900 to 1935. Bellows and Chinn (1941) have written an ex-

tensive paper on the pathogenesis of electric cataract. From the work of these and others it appears that (1) Cataract may result even when the current enters the body distal to the eye (e.g., by way of the extremities). (2) There appears to be no correlation between the degree of lens opacification and the voltage of the current.* The exact significance in this regard as to the duration of the shock which must vary in case to case is still unknown. (3) According to the cases reported a latent period extending from 1 to 19 months may precede the appearance of any lens alteration. (4) Corresponding in general to complicated cataract, and radiation cataract in particular, the lens changes occur in the anterior and posterior subcapsular regions of the cortex. Anterior capsular involvement has been noted.

Gabrielides⁴⁴² distinguishes between the lens changes resulting from high tension burns and those from lightning by the fact that in the former the opacification tends to favor the anterior subcapsular regions and the capsule itself. From the standpoint of the biomicroscopic appearance alone it is doubtful whether it would be possible, in the absence of a history of exposure, to make an etiologic diagnosis. The general form and constituent character of the opacity seem to vary according to the individual.

Koeppe⁵⁰⁷ described a case occurring in an electrician in whom contact with the right temple occurred. After three months bilateral changes in the eyes were noted. Vesicle-like protrusions of the anterior capsule and epithelium occurred, especially at the periphery. These were seen better in the zone of specular reflection and by retro-illumination. The author was reminded of keratitis vesiculosa by their appearance. The vesicles had a central silvery transparency and dark contours. Between the vesicles many smaller polymorphic opacities were present. The nucleus was more or less opacified, the

* For example, Terrien (1908) reported a case where an almost complete cataract developed in the right eye 18 weeks following a shock of 500 volts, the point of contact of the current being the right forehead. Lens opacification began after a latent period of 12 weeks. Rollet (1928) described a case which was subjected to 80,000 volts (place of contact, right parietal region). Only subcapsular opacities were found after a latent period of one month. The cataract reached maturity after 10 months. In two other instances (Koeppe, Skydsgaard) the lens opacities were more marked in the eye opposite the point of contact of the current.

posterior cortex being free of vacuoles. After one year the visual acuity became worse and the vacuoles less abundant but longer, and between them smaller opacities with angular contours were present which extended deeper into the cortex especially in the region of the sutures.

Gabrielides ⁴⁴² reported a case in an electrician who attempted suicidal electrocution. One year later the lenses in the pupillary areas appeared opaque and somewhat protruding. The opacity was irregularly polygonal, white, thick and homogeneous. Around the central opacities one could see others located immediately beneath the capsule. They were linear, white, of a silky appearance and criss-crossed indiscriminately in every direction in such a way that the whole periphery of the lens seemed to have been occupied by a spider web with small white enmeshed dots. This later appearance has also been seen by Skydsgaard ⁶¹⁰ who described a network of fine linear opacities resembling cotton fibers. Their arrangement had no relation to the lenticular structure.

Recently reports have appeared in the public press concerning the development of cataract in workers in the field of atomic energy.

Chapter Twenty-Nine

SECONDARY CATARACT

SECONDARY MEMBRANE; AFTER-CATARACT; CATARACTA SECUNDARIA *

THE term "secondary cataract" is used to describe capsular and lenticular remains as well as the products of degeneration and regeneration which are found in the wake of extracapsular cataract extraction or after spontaneous absorption that occurs following hypermaturity or traumatic perforating injuries of the lens. Besides purely lenticular derivatives in certain instances inflammatory exudates (membranes) and pigment usually derived from the iris may be included. Primarily the formation of secondary cataract depends upon the retention of the anterior capsule. The retained posterior capsule by itself (which can be seen with the biomicroscope as a delicate gossamer structure) ordinarily does not obstruct vision — although in certain instances after long periods of time it also may undergo degenerative changes and may become sufficiently opaque to require discission. However, it is when opaque anterior capsular remains adhere to the posterior capsule that we have all the necessary components of a secondary cataract. The fact that secondary cataracts and membranes are multilayered can easily be demonstrated with the biomicroscope (optic section). Frequently, these layers (anterior and posterior capsule) are separated in places by lucid or opaque material and extend parallel to one another. In such cases not only does the retained soft lens matter (cortex) become encapsulated and no longer be acted on by the lytic aqueous, but also "a partial regeneration from the capsule-containing epithelium is possible."

* For the sake of clearness this designation should not be employed to describe cataracta complicata as some writers have done.

That such regeneration occurs has been proved experimentally in animals. Clinically it is manifested in the equatorial regions by the so-called "ring of Soemmerrung."⁶¹³ Histologically this more or less circular torus-like opacity consists of remnants of retained lens matter (encysted by capsular remains) and newly formed or regenerated lens substance derived from the capsular epithelium. The character of the newly formed or regenerated lens substance varies from case to case and differs from that of normal lens fibers.

It may consist of opaque or fairly lucid material, but with the narrow beam—even when fairly clear ophthalmoscopically—it can be seen to be composed of a relucient fluid-like granular substance and hence optically is disturbing to vision * (Plate LXXVII, fig. 4).

In a sense one might be tempted to differentiate between secondary cataract and secondary membrane (Plate LXXVII, fig. 3). An important point of difference between the two is the presence of retained or regenerated lens fiber substance in secondary cataract. Clinically the distinction may be difficult to establish.

The importance of careful biomicroscopic examination of all secondary cataracts and membranes cannot be overemphasized. In Vogt's opinion, to neglect their study nowadays by means of the optic section, especially before operation, is tantamount to negligence.† It is only by such careful study that one can decide not only as to the feasibility of the type (extraction or discission) and time of operation but also as to the point where the capsule should be grasped or cut. When, soon after extracapsular extraction, free soft lens matter is present and there is no herniation of the vitreous, and according to the indications, one does not decide on immediate

* It should be noted that with time opaque material may become lucid or conversely, that following discission apparently lucid lens substance may become opaque. So that when the secondary cataract consists of any considerable amount of lucid substance it might be advisable to perform some type of extraction procedure rather than discission since this lucid material (which opacifies later) may become surrounded by vitreous—a state of affairs not conducive to absorption of the opaque lens substance.

† The experienced surgeon does not regard the operation on secondary cataract and membrane lightly. Being fully aware of the technical difficulties and complications involved he will welcome and uniformly employ this means of examination. Fortunately with the wider adoption of intracapsular extraction this troublesome chapter in ophthalmic surgery will be eliminated.

PLATE LXXVII

FIG. 1. Secondary (after) cataract with adherent iris almost secluding the pupil. The iris in the pupillary area is drawn forward and folded.

FIG. 2. Secondary membrane (after extracapsular cataract extraction) irregular pupil. Marked iris atrophy. Note cystic degeneration of the capsular epithelium and crumblike bodies.

FIG. 3. Secondary cataract (after extracapsular cataract extraction). Note irregularly round opening in the anterior capsular remains exposing the more delicate posterior capsule behind it.

FIG. 4. Secondary cataract. The area between the anterior and posterior capsular remains is occupied by cholestrin crystals.

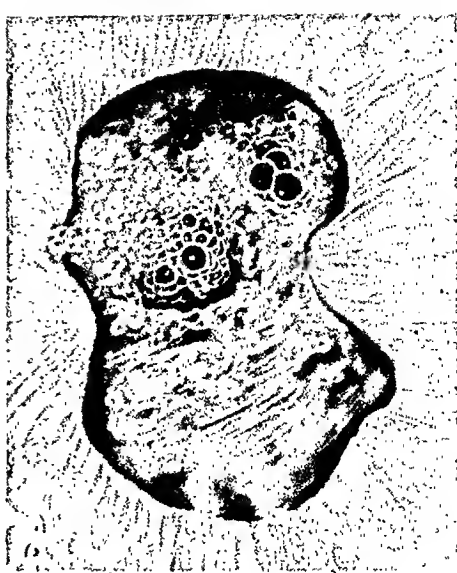
FIG. 5. Secondary cataract. Cystic degeneration of capsular epithelium (Kugeln).

FIG. 6. Secondary cataract. Cystic degeneration of the capsular epithelium. At the left the small grapelike or egg spawn cysts are viewed by light reflected from the fundus.

1



2



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3



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6



11

irrigation of the anterior chamber, the biomicroscope will permit one carefully to observe the rate of absorption and to judge the optimum time for operation. Before extraction of a secondary cataract biomicroscopy will reveal folds and thickenings in the membranes, places where proper purchase can be obtained for grasping the capsule. It will show whether iridic adhesions are present and which ones should be freed to avoid dangerous and painful traction. I once witnessed a demonstration in which almost the entire iris (atrophic ?) was pulled out with one quick jerk. This was followed by severe hemorrhage, glaucoma, and eventual loss of the eye. In cases where a discission is planned, the biomicroscope will show readily the degree of opacity (number of layers), as well as the thickness and the direction of traction bands in the secondary membrane; this will indicate the place of the incision * (usually where the membrane is thinnest), its direction (preferably at right angles to the direction of the lines of traction) and incidentally the type of cutting instrument to be employed (Ziegler, Wheeler, or de Wecker types).† Extensive synechiae and the appearance of vessels in the secondary cataract (especially the latter) should warn of the possibility of inflammatory membranes (Plate LXXVII, fig. 2). Vessels should be charted so that they can be avoided during operation, if this is at all possible, to reduce the hazard of hemorrhage. Such membranes may be very thick, and the futility of early operation upon them is well recognized. Oscillatory movements of the beam will frequently demonstrate herniation of vitreous into the anterior chamber. This is found uniformly after discission. It may appear as a definitely outlined globular mass protruding through openings in the capsule and trembling with movements of the eye. Prolapsed vitreous is always more relucient than normal, the framework being more relucient or reflecting. Fine pigment deposits are usually seen. At times after a perforating injury or needling, a tract

* After atropinization (with relaxation of zonular traction) small folds in the membrane will show the direction of the strongest traction.

† The choice of operation is governed to a large extent by the thickness of the secondary cataract. In the thin membranous types, with a Wheeler knife discission may be adequate whereas in the thicker types if extraction is not feasible a De Wecker or Berens scissors may be indicated.

of vitreous substance may be seen extending to and adherent to the incision scar in the cornea (Vol. I, Plate XXXIX, fig. 4).

It has been suggested by some (and denied by others) that anterior prolapse of the vitreous is conducive to secondary glaucoma. In the writer's experience presence of prolapsed vitreous, considering its frequent occurrence, is in itself not a *sine qua non* of secondary glaucoma. However, prolapse or anterior herniation of the vitreous into the anterior chamber may be factors in the causation of secondary glaucoma providing its adherence to the surface of the iris or posterior surface of the cornea induces peripheral anterior synechiae. Delayed reformation of the anterior chamber, incarcerated iris pillars, or the presence of an iris collar at the base of the iridectomy are contributing factors. Of no less importance is the biomicroscopic examination of the eye after discission or extraction of cataracta secundaria. This will serve not only to confirm the technical success or explain the failure of the operation, but also may help in the explanation of ensuing complications.

Clinically the biomicroscopic picture observed in cataracta secundaria varies greatly, but certain features require individual consideration. These have to do with special appearances of the capsule — e.g., iridescence, folds, and the so-called "dark slit" or "rent" line — and those changes which are associated with lenticular regeneration and degeneration — e.g., Soemmerrung's ring, Elschnig's spheres, crumblike bodies and lentoid forms, cholesterol crystals, and pigmentary deposits. Not all these changes will be found together in every case but it is not unusual for a single individual to show several of them mixed together or irregularly distributed. Except for Elschnig's spheres it is not possible to identify proliferated epithelium or the formation of new layers of hyaline capsule; these changes are seen only histologically.

Folds and Iridescence. As previously mentioned, the degree of opacification and toughness of the retained capsule (especially the anterior) varies greatly; areas of thickened and whitish capsule may be interspersed by others where the capsular rests are thin and only slightly relucet. It is not uncommon to find densely white thickened

and folded capsular remains near the margin of the iris. Directing the beam laterally to the borders of the thickened and opaque capsule and into the vitreous, the edges may be made to glow red * (Plate

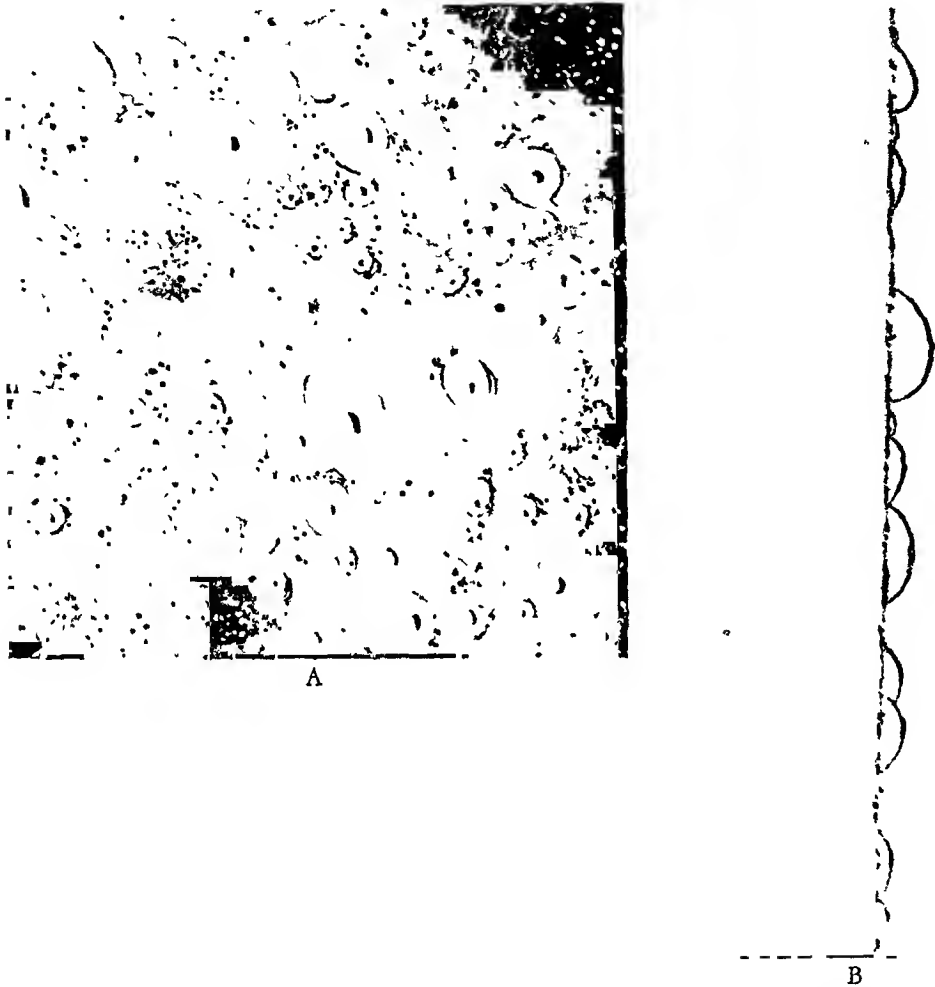


FIG. 452. Delicate (thin) secondary membrane with cystic excrescences. A. Diffuse illumination. B. Optic section. (After Vogt.)

LXXVII, fig. 6). Frequently a thinned capsular pellicle will seem to be pulled out by traction and as a result has small *radial folds*. These small folds must not be confused with the large coarser ones seen in heavy opaque membranes which are usually associated with, and caused by, the traction of firm iridic adhesions. At times some

* This phenomenon has been mentioned before, for example, in the case of the vacuoles within a cortical spoke and in the margins of ruptures of the posterior capsule. This glow is caused by reddish light (divergent rays) reflected from the fundus (as in ophthalmoscopy), and is best seen by observation along the beam's axis.

of the small folds show branchings. They are difficult to see in direct focal illumination, especially against the background of the underlying substratum. They may be more distinct by indirect illumination. The small folds in specular reflection will show double lines of reflection and when numerous, polychromatic iridescence. This color display is seen only in the mirror zone and usually is associated with the capsular shagreen. Despite this, Vogt considered that the refringence is dependent on the presence of capsular epithelium. Contrary to this conception is the fact that the color display may occur occasionally even when thick folded opaque membranes are covered by a delicate thin layer of tissue. This may suggest that the interference phenomena resulting from the diffractive effects of the multilayered membranes bring about iridescence.

When the capsule is very thin and not overly relucant, its appearance may resemble the glistening structure of the wings of the common housefly (Fig. 452 A). Slight movements of the beam or of the eye will cause changes in these reflexes indicating their optical origin (mirror zones). In Vogt's opinion, relaxation of zonular traction by atropine may cause an "ironing-out" of these folds. Graves⁴⁵⁹ also showed a similar phenomenon with secondary membranes, in accommodation studies, thus contributing further evidence in support of the Helmholtz theory of accommodation.

The Dark Slitlike Shadow Line (Vogt). One of the peculiar and still unexplained findings occasionally seen in secondary membranes is the so-called dark slit or "rent-like" line. This fine dark line (usually vertical) may be seen running more or less parallel to opaque parts of a membrane or to the border of a group of Elschnig's spheres. With widening of the angles of illumination and observation, the size of the line may change in shape; when the light is directed from the nasal side instead of the usual temporal direction it can be caused to disappear. According to Vogt, it is not a phenomenon of reflection nor can it be considered to result from a tear or rent in the membrane, because it does not change even under atropinization. If it were a rent or tear, following increased zonular traction, relaxation of the zonule with atropine should cause a change in its

appearance. It may be a shadow line from adjacent structures located at a higher level, caused partly by projection and partly by refraction. Depending on the direction of the beam a shadow effect can be elicited about single vacuoles giving the impression of circular holes in the membrane. But alteration between the angle of illumination and observation will reveal that no opening in the membrane exists.

Soemmerrung's Ring. In 1828 Soemmerrung, commenting on changes in the eye following cataract operation, described the characteristic ringlike after-cataract that now bears his name. Although this structure is seen following extracapsular extractions, it occurs most frequently following needling (in congenital cataracts or in Fukala's operation for high myopia) or after injury in which the central or axial part of the capsule is opened. If, while the central parts of the lens are being acted upon by the lytic aqueous, the margin of the anterior capsule folds or retracts and becomes adherent to the posterior capsule, all the necessary factors for the formation of Soemmerrung's rings are present (Fig. 453). Encysted between the layers of capsule there are not only the remnants of lens substance but also regenerated lens material (in the region of the nuclear equator) derived from the capsular epithelium.* Regeneration products play a role in the formation of Soemmerrung's ring; clinically cases have been observed in which the ring developed some time after discission of a pre-existing secondary membrane. It may happen that after discission, the capsular membranes curl and retract, and with release of internal tension proliferation may begin again. In such cases a large curved sausage-like structure (partial ring) may form. It is hardly likely that after a double exposure coagulated lens material could remain behind to produce such a large formation.

Histologic preparations usually show that the encapsulated material degenerates and consists of broken-down lens matter and Mor-

* Histologically it has been demonstrated also that the epithelium (similar to the corneal endothelium [see Volume I, page 424]) may proliferate and not only line the posterior capsule but also may grow around the torn edges of the anterior lens capsule and form a lining of cells on its anterior surface. These cells may secrete new hyaline material and, depending on the number of layers of epithelial cells, may form alternate layers of hyaline membranes. Between these layers varying amounts of lenticular debris and vacuoles may be found.

gagnian globules. Further degeneration may lead to calcareous and even osseous changes. The picture readily becomes apparent when the central or intervening areas of the membrane are thinned and



FIG. 453. Soemmerring type of secondary cataract (post-traumatic).

darker. The presence of such a formation may be difficult to see biomicroscopically if it is drawn peripherally behind the iris. However, in many cases maximum dilatation of the pupil will reveal it. In others the ring is more centrally located, and it rarely forms a truly complete circle but consists rather of irregularly shaped lentoid masses strung together in a ring or torus form by tenuous processes. With the narrow beam the contents of these structures may be seen to vary from a faintly relucant fluid-like substance (Morgagnian) to an intensely white, crumbly, coagulated mass. Only rarely can actual fibers be made out. Traumatic and spontaneous dislocation of

Soemmerrung's rings (after zonular degeneration) has been reported, either into the anterior chamber or backward.

Elschnig's Spheres (Kugeln), Pearls or Bladder Cells. One of the striking and frequently seen new-formations in after-cataract is the clear spheroidal (round or oval) structures first mentioned by Hirschberg (1901)⁴⁷⁴ and later more fully described by Elschnig (1911).⁴¹⁷ When massed, they have been compared to clusters of grapes, groups of small soap bubbles, or mounds of frog's spawn (Plate LXXVII, figs. 5, 6). In one case reported by Cowan and Fry (1937)³⁸⁹ a single sphere attained a diameter of 2 mm. The spheroidal bodies may appear as a flat layer of vacuoles imbedded within the membrane and seeming to interrupt it. Often the small formations may appear somewhat opaque, owing perhaps to changes in the walls or to products within the spheres. In addition, pigment in the form of fine dots or dust may be deposited on their surfaces. The larger clusters of spheres tend generally to develop on one side or the other of the membrane. They may become stationary, may disappear, or eventually may fill and occlude the pupillary opening. Cowan and McDonald (1939)³⁹⁰ estimate their frequency as 25 per cent in cases of secondary cataract. The exact origin and composition of these spheres is still unknown. Elschnig considered them to be "degeneration" products of the epithelial cells and accounted for their spherical shape by the fact that owing to the lack of normal internal lenticular tension the proliferating epithelial cells (see footnote, page 1319) assume a spherical form instead of that of a normal elongated lens fiber (Fig. 454 A, B). Because of the fact that these clusters of spheres in the beginning form in the peripheral parts, Vogt has drawn an analogy between them and the roundish equatorially located coronary opacities.

Although it is commonly accepted that these spheres are of epithelial derivation, it is possible that they may be myelin spheres derived from degenerated lens substance (cf., Cowan and Fry, 1937).³⁸⁹ As mentioned before when a considerable portion of the anterior capsule has been removed and most of the soft lens matter has either been irrigated or removed from the anterior chamber or absorbed



FIG. 454. A. Kugeln. Cystic degeneration of the capsular epithelium. (After Vogt.) B. Optic sections through the Kugeln (spheres) shown in A.

spontaneously, the posterior capsule (invisible with the ophthalmoscope) can be seen early with the biomicroscope, especially by optic section, as a delicate slightly relucet membrane. Not infrequently, even after several years, the patient will complain of fogging vision in such an eye. Biomicroscopic examination will demonstrate that the posterior capsule has become more opaque. In several cases of this kind it was possible to see a delicate vacuolar degeneration in the membrane consisting of a kind of flat layer of spheres but which were smaller than the Elschnig pearls, usually associated with anterior capsular remains. However, the possibility of backward proliferation of the anterior capsular epithelium must be kept in mind.

Crumblike Bodies and Lentoid Forms. As common as Elschnig's pearls or even more so are the so-called "crumblike" bodies, a kind of lenticular detritus (Plate LXXVII, fig. 2). These consist of small granular accumulation frequently not larger than good-sized keratic precipitates. These bodies (resorption crumbs) become free and may be found not only on the secondary membrane but also deposited on the posterior corneal surface, on the anterior surface of the iris, and in the vitreous. On the posterior surface of the cornea, such deposits may have a glasslike quality, while on the iris they stand out intensely white and granular. Recently, following an extracapsular cataract extraction two or three of these white dots could be seen with the unaided eye resting on the surface of the iris. Biomicroscopically their granular or crumblike composition and loose attachment to the iris could be established (Thiel⁶³⁶ and Riedl).

Cholesterol Crystals. Not infrequently it will be seen that the secondary membrane contains crystals, presumably cholesterol. Cholesterol has the property of crystallizing either in the form of plates of rhombic shape or as needles. These crystals may be found in small localized agglomerations in flat extended layers or as massive accumulations (Plate LXXVII, fig. 4). Depending on the illumination, they have a polychromatic luster, which is very difficult to reproduce exactly in drawings.

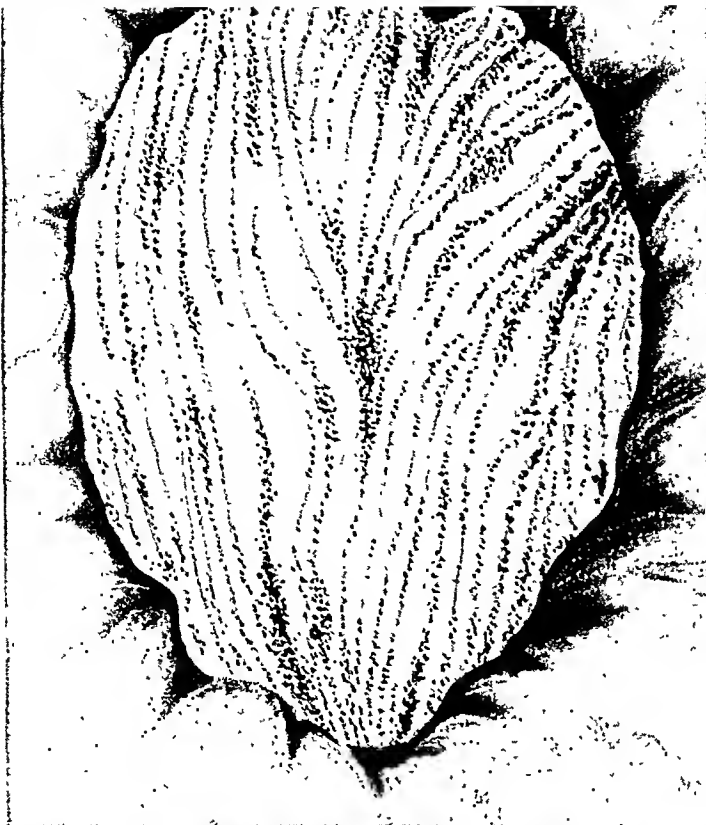


FIG. 455. Pigmented secondary cataract. (After Vogt.)

Pigmented Secondary Cataract. When pigmentation of an after-cataract is marked and becomes a dominant feature, the resulting structure is known as a "pigmented secondary cataract." The pig-

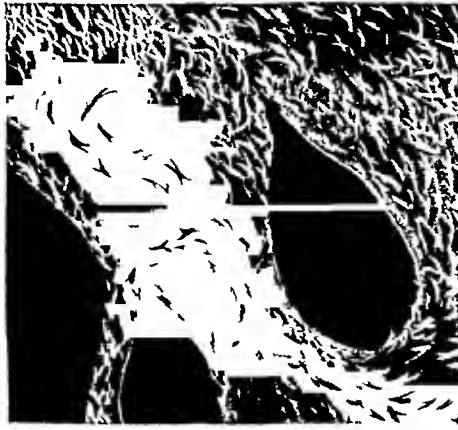


FIG. 455 (Continued).

ment may be deposited as amorphous grains or in the form of stars reminiscent of those seen congenitally in the axial region of the anterior capsule on an otherwise intact lens (Fig. 455). The stars may form a veritable net, obscuring the pellicle itself when it is delicate. The color of the pigment may vary from straw yellow to reddish-brown. It is still unknown whether the pigment is freed and deposited at the time of operation or if it migrates over the secondary membrane later. Brüchner, who held the latter impression, believed that the pigment is derived from the pigment epithelium of the ciliary body and that it migrates via the zonule to spread on the secondary membrane. The writer has seen a case in which such a possibility was suggested. Through an iris coloboma amorphous pigment ensheathing the zonular fibers was deposited wreathlike on the posterior capsule of the lens. However, since pigment is seen scattered over the anterior lens capsule so frequently (especially after iridectomy in glaucoma) a special origin and method of distribution of the pigment is hardly necessary. Amorphous iris pigment released and dispersed at the time of operative manipulation or soon afterward could be deposited and later assume its ultimate form, i.e., irregularly distributed stars with connecting processes. In this way the pigment actually takes part in the formation of after-cataract.

Occasionally, owing to iridic adhesions or even to manipulation at the time of operation, detachments of portions of the posterior pigmentary retinal layer of the iris may occur. A flattened dark-brown detached layer may be seen in the pupillary area or sometimes as an apparently structureless sheet curled up on itself. Usually one side remains attached to the posterior surface of the iris.

Chapter Thirty

THE ZONULE

GENERAL CONSIDERATIONS

LATELY, especially in view of the ever increasing popularity of the intracapsular method of cataract extraction, interest has been renewed in the morphology and dynamics of the zonular system, a structure that ordinarily does not lend itself to examination in the normal living eye. As in the lens, histologic preparations, because of technical difficulties, have failed to reveal all the details necessary to the complete understanding of zonular morphology. Owing to its delicacy, not only do fixational artefacts occur in histologic preparation but because of the long arcuate course taken by the innumerable fibers making up the zonule, it is not possible, even with serial sections cut at varying angles, to follow a single fiber from beginning to end (Fig. 456). Embryologically, the exact origin and development of the zonule is likewise still unknown. Most modern workers in this field consider that the zonule is of ectodermal origin — derived from the lens, the vitreous (tertiary), or the ciliary epithelium. Duke-Elder states: "The zonula, which is embryologically a specialized region of the vitreous, is secreted in a manner closely similar to this structure: instead of being formed from the retina itself, as is the vitreous, it is formed as an extra-cellular product of the ciliary epithelium — the prolongation of the retinal elements over the ciliary body."

Older anatomists (Petit, 1723; Zinn, 1755;⁶⁸⁶ Hanover, 1852), using teased preparations and air injections, believed that the zonule has a membranous structure.* Later histologists, using fixed prep-

* Petit believed that the zonule was formed by a portion being split off from the hyaloid membrane, thus forming a potential space between the posterior zonular membrane and the hyaloid—known as "Petit's canal." Zinn, whose name this structure commonly bears, described

arations, held that it consisted of a system of fibers which would permit of free communication with the aqueous fluid of the posterior chamber. With one or two exceptions this idea became ac-

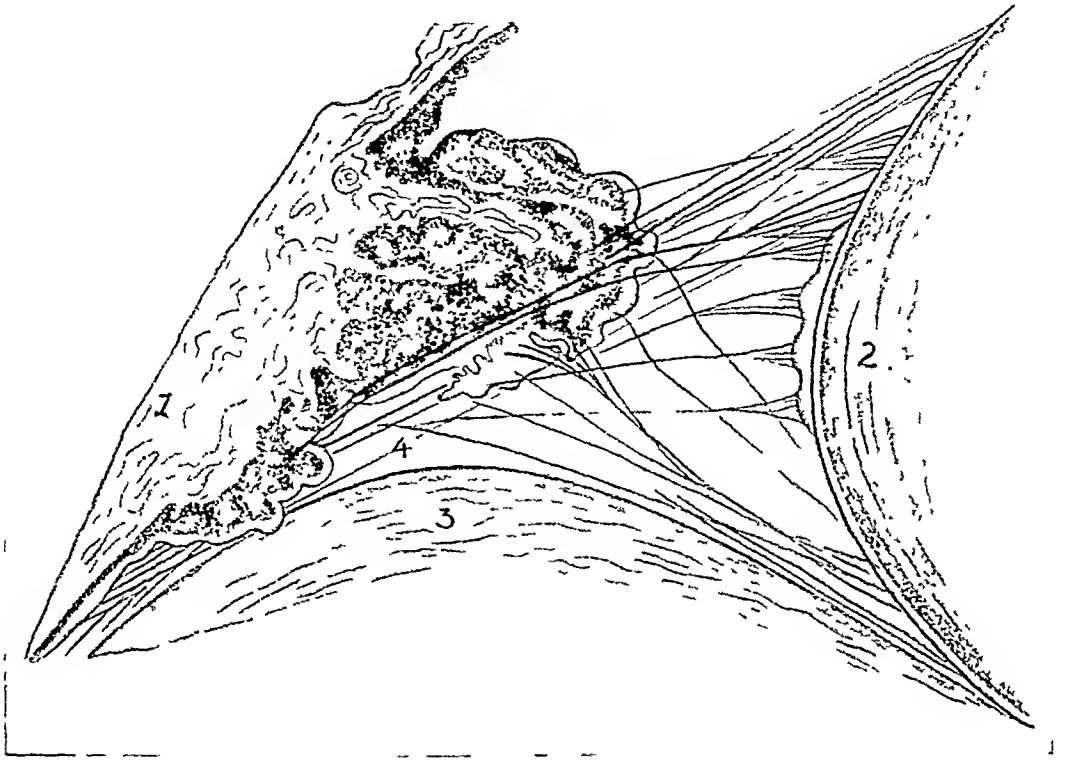


FIG. 456. Histologic section of zonule. 1. Ciliary body; 2. Lens; 3. Vitreous; 4. Zonular fibers.

cepted generally until recently. Although hinted at by Merkel (1870), the suggestion of the presence of a gelatinous cement substance between the fibers (thus reverting back to the original idea of a membrane) was not emphasized until lately.* Using fresh eyes with and without fixation, Egger (in collaboration with Vogt) sectioned them through the equator, removing the cornea, iris, and in some of the eyes, the vitreous (up to and including the hyaloid mem-

this membrane more fully as extending from the lens capsule to the ciliary body. Hanover by injecting air between the anterior and posterior leaves of the zonule demonstrated the presence of a circumlental space or canal (Hanover's canal)

* Duke-Elder states: "Embryologically, we could expect the zonule to be of the same essential structure as the vitreous and examination of the fresh eye shows a strong structural resemblance (Beauvieux, 1922; Dejean, 1928). Fibers are present but these are probably of the same nature as fibrillae of the vitreous body although more compactly aggregated together, and between them lies a clear, transparent nonstaining gel-like substance, showing the physicochemical properties of the vitreous gel."

brane). He was then able to study the fibers of the zonule *in situ* by means of the biomicroscope.* Also by means of scleral windows behind the ciliary body excellent observation could be obtained. A further development of this type of observation was made by Goldsmith (1943).⁴⁵⁴ Beside his studies of the dynamics of experimental intracapsular extraction, Goldsmith demonstrated many interesting features concerning the morphologic aspects of the suspensory ligament by the injection of dyes and air.

His findings stated, first, that the zonule is seen as a fine transparent pellicle commencing at the posterior border of the orbicularis ciliaris (flat portion) extending forward (covering the inner surface of the ciliary body) as far as the equator of the lens. This membrane consists of a system of small cords seen as fine refractile bands, especially opposite the ciliary elevations. The union of these fibers into small cords is characteristic, and two to three of them are noticed (posterior leaf) for each fissure. A unifying semisolid cement substance enmeshes the small zonular cords and transforms them into a continuous membrane (without perforations) which is extremely thin and delicate except in the portions reenforced by the zonular bundles. The posterior zonular leaf raises itself almost vertically, while the anterior zonular leaf directs itself obliquely upward and posteriorly to meet the posterior leaf at the junction of the posterior and middle thirds of the base of the ciliary processes. This continuous blended layer proceeds posteriorly, where the zonular bundles dissociate themselves into their constituent elements (zonular fibers) at different planes along their path of origin from the entire surface of the orbicularis ciliaris. The zonular fibers arise in their entirety from the complete surface of the orbicularis ciliaris. (This confirms Egger's opinion, viz., that no zonular fibers originate from the ciliary processes but that all the fibers which go to the lens have their origin further back in the region of orbicularis ciliaris-pars plana [M.L.B.].) Anteriorly the membrane is transparent and affords a good view for study of Hanover's canal (the space situated between the anterior and posterior zonular membranes). This space is filled with a viscous substance. The anterior zonular fibers which are condensed into bands do not attain the thickness of the posterior zonular bands. As soon as the anterior zonular bundles reach the ciliary processes they proceed immediately into the depths of the valleys, without preference for the lateral walls. The anterior zonular leaf is a semipermeable membrane which separates Hanover's canal from the prezonular space containing aqueous fluid.

Second, the striking anatomic point (posterior aspect) here was the complete independence of the vitreous from the lens on one hand and from the posterior zonular membrane on the other. The vitreous plays no role in the gross or micro-

* This method of examination of preparations with the biomicroscope was also advocated by Troncoso⁶⁴¹ who holds that the zonule is membranous.

scopic architecture of the zonular system—the suspensory ligament having nothing to do with the hyaloid membrane. The hyaloid possesses all the characteristics of a membrane, representing the condensation of the most superficial parallel lamellas of the vitreous.

Using fixed preparations, Egger made an exhaustive study of the arrangement and course of the zonular fibers. However, he did not stress the membranous character of the zonule nor the point emphasized by Goldsmith concerning the presence of a cement substance, which seems in the perilental area to make Hanover's canal a closed-off space. Egger did however bring out the fact (first suggested by Vogt) that only by means of specular reflection was it possible to identify the fibers themselves (when viewed from behind) in the region extending from the ciliary process to the orbicularis ciliaris (pars plana). Only by this method of illumination was it possible to recognize a fiber as it lay in front of the dark brown background. By means of a delicate glass hook it was possible to lift the fibers from the walls and valleys of the ciliary processes and to follow their course. It was seen that two groups of fibers emerged from each valley. The fibers hugged the processes* until the ends (or bases) were reached at which point they collected into two or three sagittally-ordered layers to extend to the lens. The groups of fibers on the surface of the ciliary processes were found to go toward the posterior surface of the lens. Those on the side walls, in the angle between the processes and in the valleys were directed toward the anterior surface of the lens. Between these main groups occasional fibers were found going to the equator; but this did not occur in every case (the equator may be completely free of fibers). In the latter cases all the anteriorly directed fibers were found to insert on the anterior lens surface. Here and there circular branches were seen extending between the ciliary processes. It was found that the fibers do not end at the peripheral ridges or in the valleys but that a majority almost reach the ora serrata, and that the separation of the zonule into single bundles is caused by the ciliary processes. According to measurements under high power the average length

* On the average there are about 68 ciliary processes, the distance between them being 0.7 mm.

of the fibers is from 6 to 6.5 mm. When lifting a bundle with the glass hook it was seen that numerous fibers branched singly to change their direction. In the region of the pars plica, the crossing of the radial fibers occasionally may occur. This happens when fibers which go to the anterior surface of the lens and which run over a plica (plicae ciliaris) seem to be crossed by a fiber coming from the depths of a valley which goes to the equator or posterior lens surface.

Insertion of Zonule on the Lens. From the observations of Egger and Vogt it is possible to establish that three principal insertional areas on the lens capsule can be differentiated: (1) anterior zone; (2) equatorial zone; (3) posterior zone (Fig. 456).

The anterior insertional zone according to the measurements of Egger and Vogt extends further toward the axial region (from the equator) than the posterior zone. Anteriorly this distance was 0.5 mm. and posteriorly from 0.1 to 0.2 mm. The fibrilla closest to the capsule could only be recognized in dry preparations. The course of the fibers on the lens had to be differentiated from the fine radial lines of the peripheral lens fibers whose course was almost parallel. The zonular fibers at their capsular insertions are angulated acutely as they leave the anterior and posterior lens surfaces. The divergent equatorial fibers when present pass into the circumlental space and insert irregularly at the equator as separate fibrillae. With the optic section, inspection of the circumlental area revealed two principal zones of zonular fibers (anterior and posterior) which are separated by a space (Hanover's canal) relatively poor in equatorial fibers. The width of this space in the vicinity of the fixed lens is 0.5 to 0.6 mm. Near the tips (bases) of the ciliary processes it is not more than 0.16 mm., while over the processes themselves the fibers coalesce and lie directly on the epithelium. As the fibers abut the capsular surface they form brushlike filaments, the capsular expansions of which develop a delicate membrane (the zonular lamella) (Plate LXXVIII, fig. 1). This membrane may be formed by the same cement substance that has been shown to exist between the fibers in the perilental space. The zonular lamella occupies the equatorial and periequatorial regions of the lens capsule (an area measuring

from 1.5 to 2 mm.) and hence as previously mentioned must not be confused with the more axial part that separates in senile exfoliation of the superficial capsular lamella, in glaucoma capsulare and in heat cataract. Considerable evidence, both histologic and biomicroscopic has been advanced to confirm the idea that a zonular lamella actually exists (see Plate LXXVIII, fig. 2; also Figs. 465, 466). Using special technics, its presence has been verified by Berger, Retzius and Busacca and Komai.* Clinically with the biomicroscope, Meesmann (1925) first described the appearance of a zonular lamella detached from the true capsule in a case of trauma.⁵⁴² The detached zonular lamella appeared as a circular membrane around the equator. Although normally the equatorial margin of the lens appears as a smooth regular surface actually during accommodation it is somewhat undulating (physiologic undulations or crenations).† Pathologically accentuations of these undulations can be seen biomicroscopically through a coloboma or when the lens is displaced in places where the (anterior) zonule is missing or torn away. This would suggest that the physiologic irregularities of the lens margin are related to the action of the zonule.

Other cases illustrating the existence and separation of the zonular lamella were reported by Vogt (1925),⁶⁵⁶ Stein (1926)⁶¹⁷ and Kronfeld.⁵¹⁵ Stein demonstrated a case of chalcosis in which the zonular lamella and the zonule itself was stained with a green deposit of copper. Vogt reported 2 cases (4 eyes) of detachment of the zonular

* Berger by means of maceration found that an independent lamella (zonular) could be identified in the region of the equator. With this method he was able to separate the lamella with zonular fibers attached to it. Both von Ebner⁴¹⁵ and Elschnig⁴¹⁹ presented histologic sections showing detachment of the zonular lamella. In detachment of the zonular lamella the axial region of the capsule is never affected as in senile exfoliation and in glassblowers cataract. Also in the latter conditions subluxation or luxation of the lens does not ordinarily occur. Consequently the term detachment of the zonular lamella should not be used to describe these two conditions.

† The number of these undulations or crenations which run antero-posteriorly at the equatorial edge of the lens has been estimated as about 40, corresponding to the number of ciliary valleys. Hess (1896) and later Brown (1928) showed that these indentations became more prominent after atropinization contrariwise less marked after the use of eserine. Brown demonstrated the same phenomenon in an eye (with optical iridectomy) biomicroscopically. Employing internal reflection from the contained nuclear cataract he was able to visualize the equator. While causing the patient to accommodate by fixing a near object, he saw an accentuation of the crenations and vice versa he noticed that they became less marked when a distant object was fixated.

lamella associated with bilateral spontaneous luxation of the lens.

After intracapsular cataract operation inspection reveals no zonular fibers attached to the extracted lens, perhaps the pressure exerted by the hook during the operation tears the zonular lamellae from the lens capsule rather than the fibers from the zonular lamella. However, according to biomicroscopic findings in cases of spherophakia and ectopia separation of the zonular lamella does not occur in every case. This is deduced by the frequency of the presence of torn fibers and their remnants adherent to the capsule, possibly indicating in these cases a higher degree of fragility of the zonular fibers themselves.

SITE OF ORIGIN OF THE ZONULAR FIBERS

Although it is generally agreed that most of the zonular fibers arise from the ciliary epithelium of the orbiculus ciliaris the exact way in which this occurs is still undecided (Fig. 457). V. Lenhossek (1911), in contrast suggested that their primary site of origin might be the lens, and that their ciliary attachment could be secondary. Mawas (1910),⁵³⁷ Magitot (1912),⁵³⁰ Dejean (1925),³⁹⁷ and others considered the zonular fibers of epithelial origin, arising either from the cells themselves * or between them. Terrien (1898)^{629, 630} and Wolfrum (1908)⁶⁸² held that the fibers came from between the cells originating from the underlying Bruch's membrane, or from cells corresponding to Müller's fibers (supportive tissue of the retina). Salzmann (1900) considered them to be derived from the internal limiting layer. The intimate relationship between the zonule and the internal limiting membrane (of the epithelium of the para caeca) is well demonstrated in histologic preparations. The last two conceptions would identify the zonular fibers as being of a glial nature since both the internal limiting layer and Müller's fibers are modified forms of this type of cell. If the zonule was derived from

* That epithelial cells could produce such long processes (up to 7 mm.) is not beyond possibility because, analogously, fibers of considerable length may be produced by the capsular epithelium of the lens and the fibers of the dilator iridis, from the posterior epithelium of the iris.

Bruch's membrane, it is conceivable that together with the lens capsule it is part of the glass membrane system.

In summation, it must be admitted that at present very little is

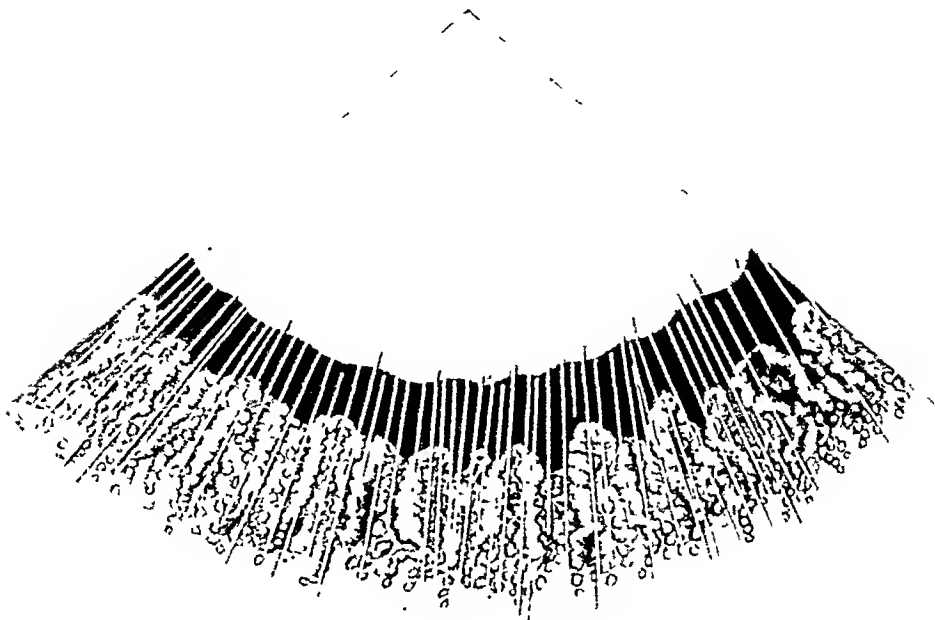


FIG. 457. Histologic section showing passage of zonular fibers between ciliary processes from the lens to the site of attachment at the flat portion of the ciliary body. (After Jess.)

known concerning the development, origin or insertion, physical (elasticity or extensibility) or chemical characteristics of the zonule. Likewise, as pointed out by Vogt, we know hardly anything about individual or senile variations and it would be of highest theoretical and practical interest to fill these gaps in our knowledge in view of the zonule's importance in accommodation and also in surgery.

TECHNIQUE FOR EXAMINATION OF THE ZONULE

In examining cases in which the zonule is sufficiently exposed so as to be accessible to the beam of the biomicroscope,* high intensity of illumination and magnification is required. The use of at least

* It is only in the presence of iris colobomas (of congenital, traumatic [ruptures and dialyses], and surgical origin) or lens displacements (subluxations and luxations) that the edge of the lens and the zonula becomes sufficiently exposed to permit adequate biomicroscopic examination. However, in the very young, when the pupil is *maximally* dilated (e.g., using adrenalin or 10 per cent neosynepherin, the insertion zone of the anterior zonula fibers to the (normally placed) lens may just be visible. With the exception of the directly traumatic cases, lens displacements may be hereditary or congenital or may arise spontaneously usually secondarily to high grade congenital myopia, hydrophthalmos, inflammations (iridocyclitis or uveitis) and cataract (particularly in shrunken lenses).

40 \times magnification is desirable. As is the case of the contents of the anterior chamber and the vitreous where the delicate structures (invisible by diffuse illumination) can only become visible by means

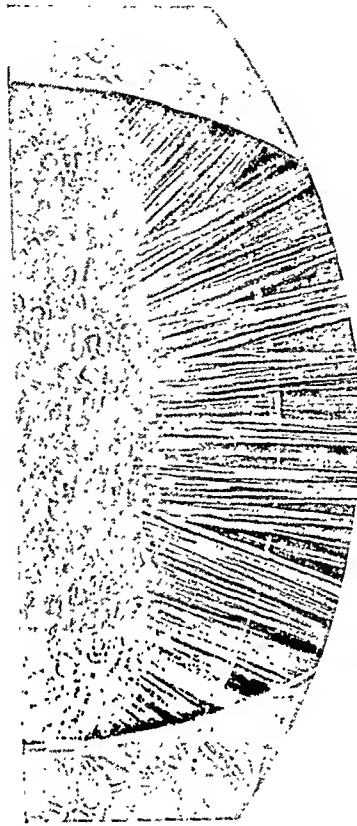
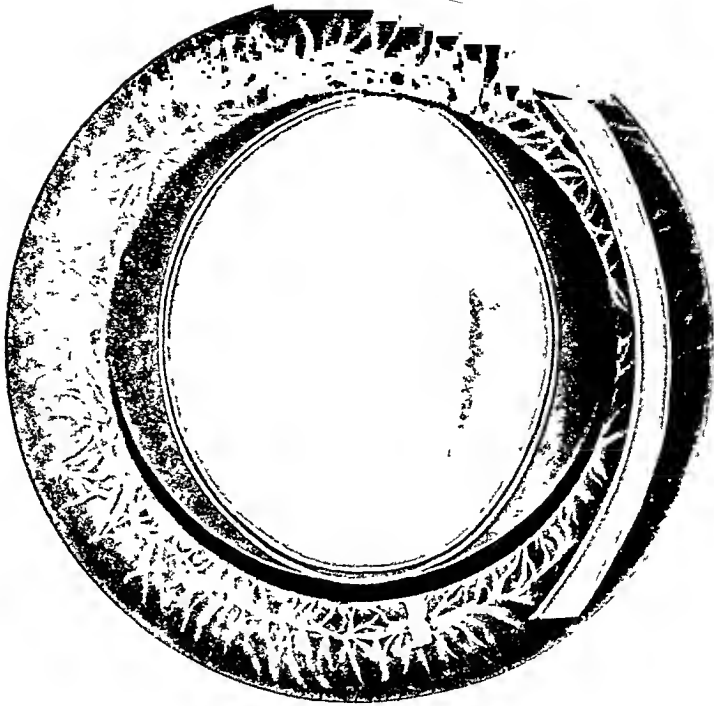


FIG. 458. Examination of the zonular fibers by means of specular reflection. Note shagreen of the anterior capsule. Posterior zonular fibers are seen faintly behind the stronger and better developed anterior ones.

of the tyndall phenomenon, all examinations of the zonule must be performed by direct focal illumination. Similarly, appearances and visibility may alter with the direction of the light. Therefore when examining the zonule the beam should be directed first from the temporal side and then from the nasal. Preferably the examination room should be dark and the examiner at least partially dark-adapted. When feasible the pupil should be dilated. Both the wide beam and optic section should be employed. Since the intensity of illumination is decreased with the narrow beam, overloading the light source will give added brightness. By means of specular reflection it is often possible to render visible zonular fibers which may be hardly recognizable in ordinary direct focal light (Fig..458). Slight



A



B

FIG. 459. Spherophakia (ball-shaped lens). A. Low power. Note reflex from equator. B. High power. Note absence of peripheral divergence of outer zones of discontinuity. (Modified after Vogt.)

(but slow) oscillatory movements of the beam are frequently of great help in accentuating visibility. This will be understandable when one considers that the zonular fibers are prismatic in shape rather than round and hence their reflection (and visibility) will vary with the direction of the light. On the other hand although the visibility of the fibers themselves become enhanced by specular reflection, those located deeper may be obscured in the reflex of the more anterior and hence details concerning the deeper ones (although not brilliant) will be seen better in the beam outside the mirror zones. Even in the most favorable cases only that part of the zonule which extends from the lens to the tips of the ciliary processes will be seen. In directing the beam obliquely the margin (equator) of the lens becomes highly dispersive, appearing as a white line (see Fig. 459). The fibers going to the anterior lens capsule will be seen passing anterior to this line to their point of insertion. In addition, when the lens is displaced (as in ectopia), passage of the beam through it at some distance from the subluxated edge will cause this edge (equator) to glow as a reddish line. This is the result of internal reflection, a form of "lens scatter." The fibers themselves appear as delicate threads that glisten when viewed by specular reflection. They have a tendency to form groups and bundles; this is not surprising since, as already indicated, the fibers coming from the valleys are necessarily collected in bundles because of the ciliary processes.

The beginner is often astonished to find how far forward (axial to the equator) the fibers are really inserted. Posteriorly the fibers insert closer to the equator. Anteriorly as well as posteriorly many of the fibers are collected in bundles. In most instances in which it becomes possible to see the fibers, it is rare indeed to find them entirely intact. In certain areas the bundles may be found to be spread apart or missing (Fig. 462; Plate LXXVIII, figs. 2, 5, 6). As a rule those that we do see are stretched out. Frequently remnants of torn fibers will be seen curled up at their attachment to the capsule. In others all that remains at the site of the capsular detachment and the fibers are small white dots (Plate LXXVIII, fig. 1). In prac-

tically all of the cases in which the fibers can be seen (since some abnormality or pathologic process has made this possible) they are already changed. The changes consist of deposits of white and brown (pigment) dots between or on the fibers (Plate LXXVIII, figs. 1-6). Often when they are profuse, such deposits may form a sheathing about the the fibers for varying distances. Such changes probably precede fragility and enhance the visibility of the fibers. Erggelet⁴²¹ was the first to describe these alterations. In one case, after iridocyclitis, it appeared as if the fibers were covered with dust. Meesmann⁵⁴⁰ considered increased opacification, swelling, and deposits as signs of degeneration of the fibers. Comberg³⁸⁷ (cited by Vogt) described deposits of white and red blood corpuscles on the fibers as well as fibrinous agglutination of the fibers themselves.

As previously mentioned, there is still a great paucity of knowledge concerning the tensile strength and elasticity of the zonular fibers. That these fibers can undergo considerable stretching without being entirely torn off is apparent in cases of subluxation and even of total luxation. Often when the lens is completely luxated into the anterior chamber, it is still possible to see stretched or broken zonular fibers attached to it. That the fibers can stretch for several millimeters and remain so for long periods before rupturing is evidenced in cases of pathologically shrunk lenses. Whether or not it is possible for detachment of the ciliary epithelium or the layer below it (Bruch's membrane) to occur in the region of the orbiculus ciliaris is not certain. It is more likely that in an acute (e.g., trauma) or chronic overstretching (spontaneous luxation) that the zonule is torn from its lens attachment rather than from its point of origin. As Vogt has pointed out, some authors have believed that in cases of congenital ectasia, spherophakia, and microphakia the fault lay in abnormally long fibers, genetically determined. He doubted this hypothesis because in all his cases in which anomalies and long fibers occurred, torn or broken fibers with pigmented or whitish deposits were found. These abnormalities do not result from otherwise normal congenitally long fibers but rather from an inherent inferiority or weakness of the fibers themselves. In other words, in

these conditions it seems that we are dealing with a rudimentary zonule which undergoes degeneration owing to a faulty or defective anlage which is conducive to stretching and tearing of the fibers.*

THE ZONULE IN CERTAIN HEREDITARY AND CONGENITAL ANOMALIES

In certain ocular defects, the zonule or parts of it may be accessible to biomicroscopic inspection; the zonule in congenital coloboma of the iris and lens and in ectopia lentis, in spherophakia, and in microphakia is therefore considered in this section. Some writers consider that all these conditions are related in so far as they result from varying degrees of zonular fiber destruction. That is, with partial involvement ectopia will result and that when all the fibers are stretched (loose or torn) or aplastic, microphakia or spherophakia may be induced. Microphakic (congenitally small) lenses are always ball-shaped or spherical. Therefore the difference, if any, between the terms microphakia and spherophakia can only reside in the size of the lens. If it is true (as seems likely) that all these anomalies are the result of a hereditary defect in the anlage of the zonule then we might consider that ectopia, spherophakia, and microphakia simply represent successive degrees of deterioration. In microphakia and spherophakia, the lens is in a perpetual state of complete accommodation; being rounded, it approaches in shape that of the fetus or newborn and resembles the lens of young children in the state of total accommodation. Thus it will be found that considerable lenticular myopia is present even when corneal curvature and axial length of the eyeball are normal.

THE ZONULE IN CONGENITAL COLOBOMA OF IRIS AND LENS

In colobomas of the iris the edge (equator) of the lens will lie behind the pillars and run at right angles to them. The edge of the lens may be straight, crenated or slightly undulating (Plate XLVI,

* Besides the hypothesis of congenitally long fibers for the causation of microphakia (Saeger and H. Meyer) Marchesani has suggested that the weakness of the zonule may result from a hyperplasia of the ciliary body. In addition to a defective zonule Gnad considered the possibility of an abnormally small lens anlage.

fig. 6). If one angulates the eye or the beam so that the mirror reflex approaches the lens equator the bundles of zonular fibers will readily become apparent. Using the optic section and high power, $40\times$ or more, and by alternate changes in the direction of illumination with corresponding changes in the focus of the microscope, one will be able to distinguish between the anterior and posterior bundles of the zonular fibers (Fig. 458). It will be seen in many cases that some of the anterior fibers are wholly or partially missing or torn. In the latter case remnants of torn fibers will appear as white dots on the lens capsule or as short curly threads adhering to it. These may be found as far as one-half the distance between the equator and the axial region of the lens. Ordinarily the posterior fibers are not inserted as close to the axis as the anterior. Being shorter, the posterior fibers are straighter, and unless the lens is subluxated they are less liable to be torn. Small white points or pigment granules may be found to be deposited on or between the fibers, particularly when cataract is present or when the lens is displaced (Plate LXXVIII, fig. 6). In those cases in which there is considerable tearing or in which the anterior zonular fibers are missing, the anterior surface of the lens may be more curved than is normal.* Probably from the same cause, the degree of peripheral divergence of the anterior zones of discontinuity may be less than that of the posterior. When larger openings in the anterior zonular bundle are associated with similar defects in the fibers directed to the posterior capsule, a forward herniation of the vitreous may occur. In such instances the surface of the herniated vitreous may be accented by deposited pigment cells. The association of ectopia lentis with iris colobomas is well known. Retractions of the lens margins may be considered as colobomas of the lens. Such an effect can also be produced by a contusion; following tearing of the zonule a localized retraction and rounding of the equator may occur (capsule remaining intact) (Fig. 464). Likewise, undulations of this type may be seen in cases of shrunken lenses (which are usually somewhat luxated or ectopic) especially after an

* This again emphasizes the importance of the anterior zonular fibers in the process of accommodation since it is chiefly in anterior parts of the lens that relaxation occurs.

iridectomy has been performed in the region of the ectopia (Fig. 463). When these localized undulations of the lens margins are marked, they may resemble "pseudopodia" (Vogt). In studying the zonular attachments in these instances it will be seen that the anterior zonular fibers are found only in the region of the pseudopodia and not in the retracted areas between them. Probably, this is an indication that the stronger anterior fibers attached to these processes contributed to their formation owing to the lack of traction in the retracted areas. Evidently, in the absence of zonular traction the lens margin (equator) retracts and becomes more rounded sagittally. It is interesting to observe that the posterior zonular fibers may still be present even in areas of retraction between the "pseudopodia."

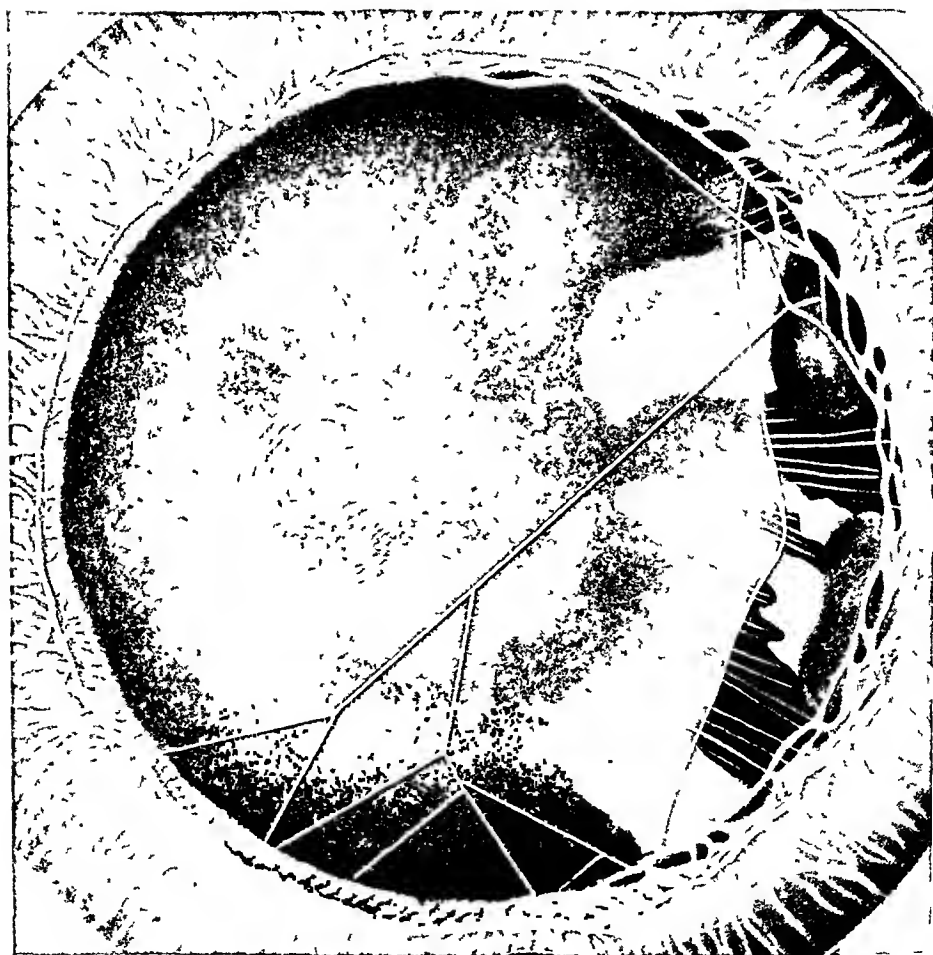
HEREDITARY SPHEROPHAKIA ("BALL-SHAPED LENS,"
LENS ROTUNDA AND MICROPHAKIA)

In microphakia (congenital smallness of the lens) the lens itself is always round or spherical (spherophakia) (Fig. 459). The equatorial diameter is thereby reduced. In one of Vogt's cases it was 7.5 mm., a figure corresponding to that given for microphakic lenses by Fleischer and Saeger.⁵⁴⁹ Mayer and Holstein found more than 20 cases of microphakia reported in the literature. In only one of these was the condition limited to one eye. In several it was found in siblings of consanguinous parents, and hence it is probably transmitted as a recessive character.* Vogt has estimated that in his cases the sagittal thickness of the lens was one-quarter greater than in the normal lens. This accounts for the high degree of lenticular myopia even when the corneal curvature and axial length of the eye are normal. This was verified by the fact that in one case, before lens extraction there was a myopia of 12 diopters, while after lens extraction in the same eye a hyperopia of 12 diopters was found. In microphakia or spherophakia the peripheral divergence of the zones

* In all the hereditary diseases of the eye there is perhaps not one degenerative change that cannot first appear in later life (abiotrophy) as well as congenitally. This may occur in the zonule (Vogt) in a way similar to senile degenerations of the lens and retina, and may account for the fragility of the zonules (as seen surgically in intracapsular cataract operations) even as a presenile manifestation, as well as in old age.



A



B

FIG 460 A Shrunken lens in lower part of anterior chamber Loosened capsule is folded (After Bonnocolto) B Dislocated lens with pupillary membranes (After Bonnocolto)

of discontinuity is diminished to a high degree, thus suggesting that this phenomenon is related to zonular traction. As discussed in the chapter on the normal anatomy of the lens (pages 1016-1017),

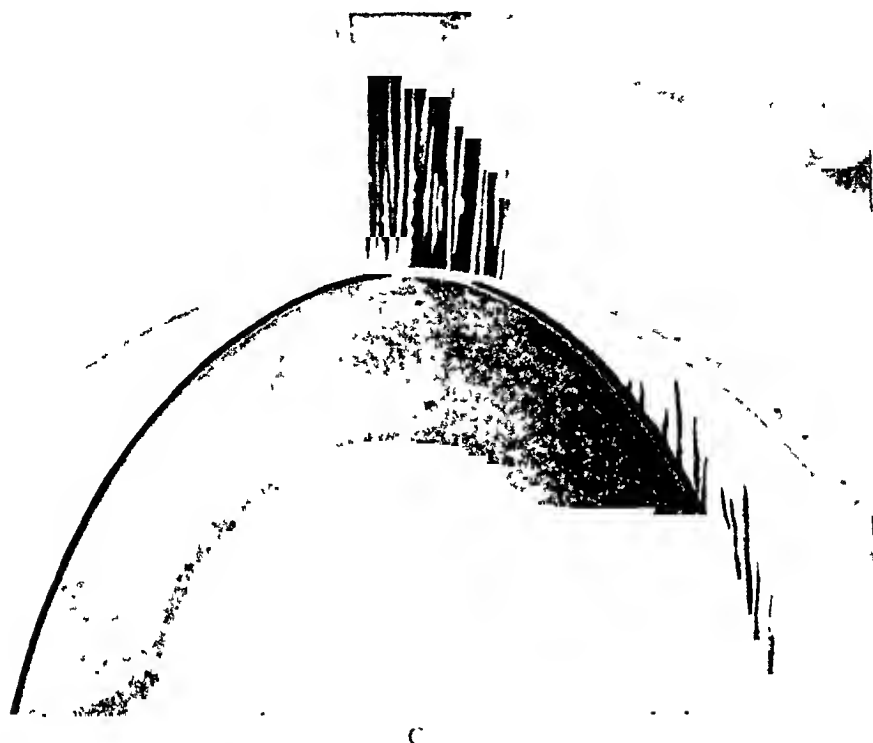


FIG. 460. (*Continued*) c. Ectopia lentis. Note absence of zonular fibers excepting for a few above and torn remnants on anterior capsule; also reflex line from the posterior capsule

zonular traction plays a role in the formation and development of the suture system — which in turn permits dorsoventral flattening of the lens. Normally and especially after the adult nucleus is well delineated, it will be seen that equatorially its surfaces angulate acutely (page 998). This may also result from zonular traction since in spherophakia, in which zonular traction is missing, the equator of the nucleus is circular.

As already indicated the finding of torn or absent zonular fibers, with or without pigment deposits, strengthened the concept that microphakia, spherophakia, and ectopia result from a congenital weakness or aplasia of these fibers. This would tend to refute the view generally held that these conditions result primarily from a congenital abnormal lengthening of otherwise normal fibers, which with consequent lessened traction permit the lens to maintain its fetal form.

When the anlage of the zonule is completely defective, varying degrees of microphakia result. When the defect is localized to one zone, ectopia follows. In other words, these anomalies may arise from an inherited weak zonule, the lens itself only being secondarily affected.

Owing to unequal traction of the remaining pathologic fibers, the small, round or spherical lens may be ectopic; or if it is supported within the pupillary area, it may prolapse somewhat into the anterior chamber. As a result, the anterior chamber may be very shallow, especially axially. The surface of the lens in this case more or less parallels the curve of the cornea. When the lens does not protrude into the pupil, the depth of the anterior chamber may be increased. Herniations of the vitreous are not uncommon. If the lens protrudes forward into the pupil, the use of miotics may further accentuate the ball valve action and may cause a rise in intra-ocular pressure. Conversely, mydriasis may cause a reduction of the pressure. Complete luxation of the lens into the anterior chamber with secondary glaucoma is a complication common to this condition and to ectopia lentis as well. This may also occur when the lens is shrunken (Fig. 460 A). Notwithstanding their congenital nature, luxations may be brought about by progressive zonular deterioration abetted mechanically by sudden strain incident to external trauma, or even after coughing, sneezing, or exercise. Although these conditions (microphakia, spherophakia, and ectopia lentis) are congenital, they may not remain stationary. Progression of zonular deterioration may lead to further lenticular displacement. Owing to the poor vision, many of these eyes develop varying degrees of nystagmus. Iridodonesis as well as phakodonesis is present.

Biomicroscopic examination in these cases should be done with the pupil dilated, if possible. As the beam is passed through the lens, a linear reflex may be seen derived from the posterior capsule. The reflex becomes longer and more distinct as the equator is approached (Fig. 460 C). Parallaxically the reflex moves in a direction opposite to that of the beam. The condition of the zonular fibers themselves will vary from case to case. They may be entirely absent (rare) or

the bundles may be, as is more likely, missing in certain areas. When present they show varying degrees of pathologic change and are seen best by specular reflection. Usually their insertion is more peripheral than in the normal eye. Curled remnants of torn fibers may be found attached to the lens, or as small white dots at the site of their former attachments peripherally on the anterior capsule. Characteristically the fibers present are delicate, loose and wavy as compared to normal ones. In many cases pigmented or whitish deposits will be found deposited on the fibers or between them. In places, these deposits may be so thick as to form a coating similar to the insulation on a wire. (See Plate LXXVIII, figs. 3, 4, 6.)

HEREDITARY AND CONGENITAL ECTOPIA LENTIS

Hereditary and congenital displacement or subluxation of the lens must be differentiated from the luxations following trauma and those appearing spontaneously subsequent to intra-ocular disease. In all, the displacement of the lens results from a damaged zonular apparatus. Hence from a purely biomicroscopic viewpoint (not considering the history and damage to the neighboring structures which usually occurs in trauma) the appearance of traumatic ectopia lentis does not differ markedly in itself from that seen in spontaneous hereditary displacements (lens sizes being equal). However, in hereditary and congenital ectopia, the condition is practically always bilateral and symmetrical, the displacement being predominantly upward or upward and outward, and is found in successive generations, transmitted usually as a recessive character although it may be dominant (Waardenburg). It may occur alone (simple form) unaccompanied by any other anomaly (ocular or otherwise); it may also occur with other ocular defects (iris coloboma, pupillary membranes or lens opacities) in the absence of systemic defects, or it may be part of a syndrome, in which it is associated with widely disseminated abnormalities of the bones and muscles, especially of the extremities, e.g., Marfan's syndrome (arachnodactyly and dystrophia adiposa congenita). In ectopia lentis the partially displaced

lens may be stationary or it may become completely luxated, gradually or suddenly. When the increase in displacement is gradual, progressive destruction of the zonule may be suspected; if it is sudden, trauma (even when indirect, e.g., sneezing or bending) must be considered as a possibility. As already noted, the complication of luxation of the lens into the anterior chamber leads not only to secondary glaucoma but also to opacification of the cornea, provided the lens remains in contact with it.

Excepting for the hereditary nature of this form of ectopia very little is known about its exact pathogenesis and consequently numerous theories have been advanced to explain it.* However, owing to the fact that in no case examined biomicroscopically have perfectly normal fibers been found (Vogt) it would seem that the most probable explanation lies in a defective anlage of the zonule. When this is complete or nearly so, spherophakia results with partial ectopia. As is the case with many other hereditary degenerations, the effects are not manifest at birth but may appear later or even as a presenile alteration (abiotrophy). Hence it is not surprising that this condition, besides being congenital, can suddenly appear at any time after birth and like other abiotrophic degenerations (presenile or senile macular degeneration or even senile cataract and graying of the hair) may be progressive. The most frequent complication in ectopia is complete luxation and its attendant sequelae (prolapse of the lens into the anterior chamber followed by iridocyclitis, keratitis and secondary glaucoma). Posterior displacement of the lens into the vitreous may not be accompanied by untoward complications or increased intra-ocular pressure. Vogt (1905 and 1925) reported a pedigree extending over 100 years where without any known cause (abiotrophy ?) 16 out of 46 descendents were affected. Between the

* Among these are: that like lens colobomas, ectopia may result from the persistence of embryonal vessels (Hess, 1911; Rones, 1934 and others); abnormality in closure of the fetal cleft interfering with development of the zonule; high grade myopia causing a disproportion between the size of the lens and the space allowed for it could result in stretching and tearing of the zonule; eccentric location of the developing lens; abnormally long but otherwise normal zonular fibers; also because of the occasional finding of pupillary membranes and strands, (some of which come forward from behind the pupil and lie on the anterior surface of the lens border) it is thought that traction may be exerted. (Fig. 151b)

ages of 20 and 70 years these persons, whose eyes previously were normal, began to have trouble with their vision. The lens began to slip down, and the visible zonule above was seen to be extended,

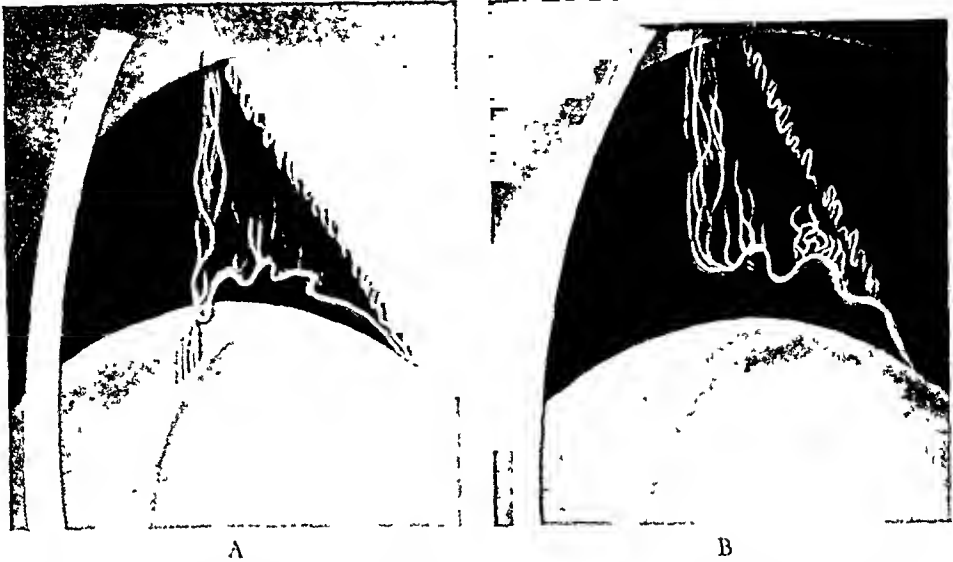


FIG. 461. Hereditary ectopia lentis. A. Detachment of the zonular lamella. (After Vogt.)
B. Complete separation of the zonular lamella anteriorly from the capsule. (After Vogt.)

fragile and torn. Especially interesting was the fact that separation of the zonular lamella occurred in some instances (Fig. 461 A, B). Eventually many of these patients developed glaucoma, some with and others without total luxation.

According to the conception of an inherited weak zonule the changes in the lens itself in spherophakia and ectopia are secondary. Pathologically many observers (Seefelder, Fuchs, Reese and Clark) have noted the presence of poorly developed and retracted ciliary processes. The significance of this in ectopia is not completely understood but most likely it is secondary to imperfect zonular development.

From the standpoint of biomicroscopy the findings in ectopia will vary somewhat with the degree of lenticular displacement. The lens may be displaced so far that its edge cannot be seen in the undilated pupillary area (complete aphakia). In this case, passage of the beam will reveal the undulating surface of vitreous body *in situ* behind the pupil. Occasionally herniations of vitreous anteriorly may occur. In

such instances of marked displacement it is rare to find many zonular fibers in the area of the small pupil. Iridodonesis is usually present. The vitreous generally shows changes in its trabecular structure and also may contain deposits. With dilatation of the pupil — always difficult to obtain in ectopia — the margin of the lens may come into view.* However, in this type of case, unless the ectopia is associated with iris coloboma (a not too infrequent combination), it is often impossible with the biomicroscope to see the margin of the lens (when displaced inferiorly), because the rigidity of the apparatus does not permit sufficient angulation of the beam. When this is so, it is necessary to resort to the ophthalmoscope, which usually will reveal the edge of lens as a dark rim across the red fundal reflex. When the lens is not overly displaced, passage of the focal beam will readily reveal its presence and its margin. If the beam is directed so that it passes partly through the lens and partly into the aphakic part of the pupil, an abrupt and sharp contrast will be seen between the pearly gray reluctance of the lens and the dark aphakic portion. Opacification of the lens will enhance the contrast. By means of internal dispersion, light from the beam (a kind of scatter) causes the margin of the displaced lens to glow as a bright reflex. As in spherophakia the zonular fiber bundles in the area of the defect may be totally or partially missing. The fibers may be stretched or torn, in which case remnants and rests may be found at the site of their former attachment. But in certain cases exaggerated branching of the fibers (to form brush-like endings at the lens) may be very marked. The fibers begin to branch and diverge at a greater distance from the lens than is normal. Pigment and whitish deposits are commonly seen on or between the fibers. There may be a definite decrease in peripheral divergence of the zones of discontinuity and a rounding of the equator of the adult nucleus (Fig. 459). With luxation of the lens into the anterior chamber, the edge of the lens may be seen to ride over the pupillary margin. This may cause the formation of a concentric shadow line on the iris near the pupillary margin when the beam is

* The difficulty in obtaining mydriasis in all forms of ectopia is well known. In a case of Marfan's syndrome, Theobald found histologically that the dilator iridis was not developed.

directed over the lens margin (Vogt). Usually only delicate and retracted remnants of the zonule (attached to the lens) will be seen. Detachment of the zonular lamella of the lens capsule may occur in hereditary ectopia and spherophakia as well as following trauma. The separation is usually irregular and may be restricted to one or more zones. It appears as a delicate membrane separated from the underlying equatorial capsule by a lucid interval (Fig. 461 A). Depending on the traction of the remaining zonular fibers, its surface may be smooth or undulating. Since the anterior fibers of the zonule are the strongest, the zonular lamella may be torn from the anterior capsule, and its free edge may be folded forward. This is exemplified by the frequency with which the anterior zonular fibers remain attached to the separated lamella (Fig. 461 B). These fibers are seen to be loosened and wavy. Posteriorly, although it is separated up to its pre-equatorial extremity, the lamella does not so readily tear away entirely from the capsule. This could indicate that the weaker equatorial and posterior zonular fibers tear off more easily from the lamella than does the latter from the capsule. The attachment of the lamella to the capsule thus appears to be stronger posteriorly.

The association of ectopia lentis with generalized abnormality of the musculature and bones (arachnodactyly) in Marfan's syndrome * is an example of a hereditary condition in which generalized mesodermal and ectodermal defects are associated, e.g., the dilator pupillae and zonule (nowadays the zonule is regarded as being of ectodermal origin). Evidently such a combination illustrates the genetic phenomenon of crossing over and then the coupling of

* According to Weve about 40 per cent of those having arachnodactyly have ectopia lentis. The extraocular findings in Marfan's syndrome which have been noted (not all being found in any single case) by numerous authors and cited by Weve, Vogt and Rados are: abnormal thinness of the extremities, the elongated phalanges (spider fingers) and contractures of their joints (hence the name "arachnodactyly"), small chest, winged scapulae, spinal curvatures, high arched palate, deep seated eyes, contraction of articulations, spur of calcaneus, duck-foot, flat feet, long and thin face (dolichocephalic), double row of teeth, enlargement of sella turcica, valvular heart disease, patent foramen ovale, large ear lobes, reduced number in the lobes of lung, abnormally long intestine. In the eye, in addition to ectopia lentis and spherophakia, there may be incomplete development of the iris stroma and owing to defects in the sphincter iridis and especially in the dilator iridis (both of ectodermal origin) dilatation of the pupil may be accomplished with difficulty.

single characters or defective hereditary qualities regularly within a single chromosome. In this way characters which somatically are far apart may be closely linked.*

The frequent association of ocular anomalies and those of the extremities is well known. Recently I reported two cases in which pupillary membranes were associated with syndactyly. According to Sorsby⁶¹⁴ the fact that in the segmented animal the eye and tentacles are homologous, being the sense organs of different segments, may have some significance in this connection. There is now evidence to support the view that the inheritance of Marfan's syndrome may have a recessive as well as a dominant character. As in simple spherophakia and ectopia, monocular as well as binocular, spontaneous total luxation of the lens may occur either into the anterior chamber or vitreous. This has raised the question as to whether Marfan's disease is progressive. Progression in the strict sense as seen in dystrophies may in hereditary conditions only be a manifestation as Bellows states of "increased susceptibility to injury and inflammation."

Biomicroscopically, except for the possibility of detecting defects in the stroma of the iris, our interest lies in the accompanying ectopia lentis. Since this differs in no way morphologically from simple ectopia just discussed, no further description is necessary. However, those who are interested in a more complete survey and discussion of this syndrome are referred to the excellent papers by Weve and Rados.

PATHOLOGIC ALTERATIONS OF THE ZONULE FOLLOWING INFLAMMATION

Alterations in the zonule after inflammation, i.e., iridocyclitis, may be observed biomicroscopically in the presence of coloboma of

* In addition to the combination of ectopia-myopia-heart disease (Strebel-Steiger), Vogt lists several combinations or complexes as examples of the genetic phenomenon of crossing over and coupling of characters so that in the end they become linked inside a single chromosome—and later appear as well defined syndromes. Among these are retinitis pigmentosa and deafness; axial myopia with large interpupillary distance (large skull base); axial hyperopia with small interpupillary distance; aniridia with aplasia of the macula, sex-linked recessive aplasia of the macula with corneal astigmatism; corneal astigmatism with albinism; syndrome of blue scleras, deafness and fragility of the bones; there is also statistical evidence to prove the frequency of hyperopia in morons and myopia in intellectuals.

the iris (either of congenital, operative or traumatic origin) or when shrinkage of the lens permits inspection. Not unlike the findings in hereditary alterations, the zonule shows varying degrees of destruction. The fibers may be torn or absent in places. Those that are seen, depending on the degree of traction, are frequently stretched and taut. Others may be coiled, and torn remnants may be seen attached to the lens. In some cases because of increased reflection of the anterior layer of fibers, repeated changes in the direction of illumination will be necessary to bring out the lesions in the deeper fiber bundles. Specular reflexes which enhance the visibility of the superficial fibers may conceal torn and coiled fibers behind them. When all the fibers are missing in a certain sector, irregularities or notching (coloboma) of the corresponding segment of the lens margin may be seen (page 1341). This is more likely to occur when the lens is cataractous and shrunken.

A very pronounced characteristic change after inflammation is the sheathing of the fibers by pigmented and whitish deposits (Plate LXXVIII, figs. 1, 3, 4, 5). This sheathing may be continuous or interrupted and causes the fibers to stand out in bold relief. In cases of old iridocyclitis, heavily pigmented fibers appear like miniature brown cords of rock candy. These changes are symptoms of zonular destruction and explain the ease — well known to surgeons — with which complicated cataract (so often a sequelae of chronic iridocyclitis) can be extracted intracapsularly. After freeing any iridic adhesions, only slight pressure is required to break the fragile zonular fibers. This also holds true for shrunken cataractous lenses resulting from inflammation, trauma, or hypermaturity (senile cataract) (Figs. 462 and 463). In the latter instance, complications such as iritis and secondary glaucoma are not uncommon (page 1139). The high degree of fragility of the zonule explains the spontaneous luxation that may occur in cases of cataracta complicata and shrunken lens. Phakodonesis as well as iridodonesis usually are demonstrable.

In addition, in cases of shrunken lenses in iridocyclitis with operative colobomas, Vogt has described the parallel striping seen by specular reflection on the anterior surface of the vitreous (Fig. 464).

PLATE LXXVIII

FIG. 1. Subluxated lens. Chronic glaucoma. Almost total iridectomy. Remains of iris attached to the posterior corneal surface. Note that the zonular fibers pass through the circumlental area and extend between the tips of the ciliary processes.

FIG. 2. Shrunk cataractous lens. Iridectomy above. Sparse zonular fibers seen attached to the lens. Questionable separation of the zonular lamella.

FIG. 3. Shrunk cataractous lens. After iridocyclitis. Iridectomy. Note pigment deposits on the zonular fibers. (After Duverger and Velter.)

FIG. 4. Subluxated lens. Shrunk and cataractous. Chronic iridocyclitis. Note pigment deposits on the zonular fibers and on the posterior lens capsule at their attachment.

FIG. 5. Subluxated cataractous lens after iridocyclitis. Zonular fibers are sparse in number. Some of them show fine deposits.

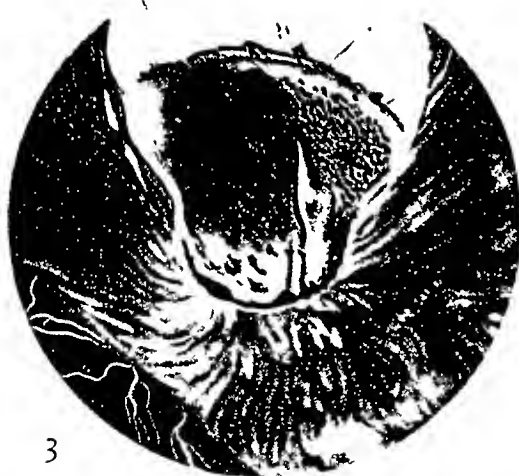
FIG. 6. Aniridia. Subluxated lens. White deposits on zonular fibers. Lamella type of lens opacity.



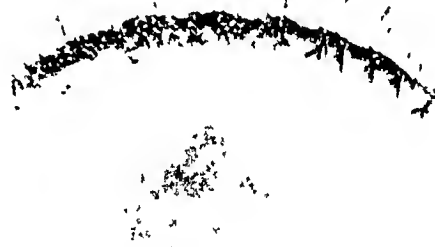
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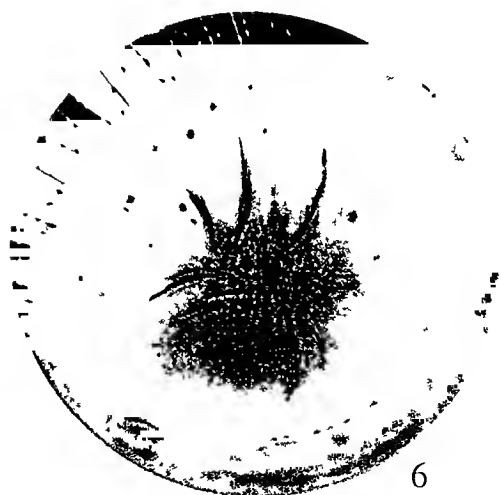
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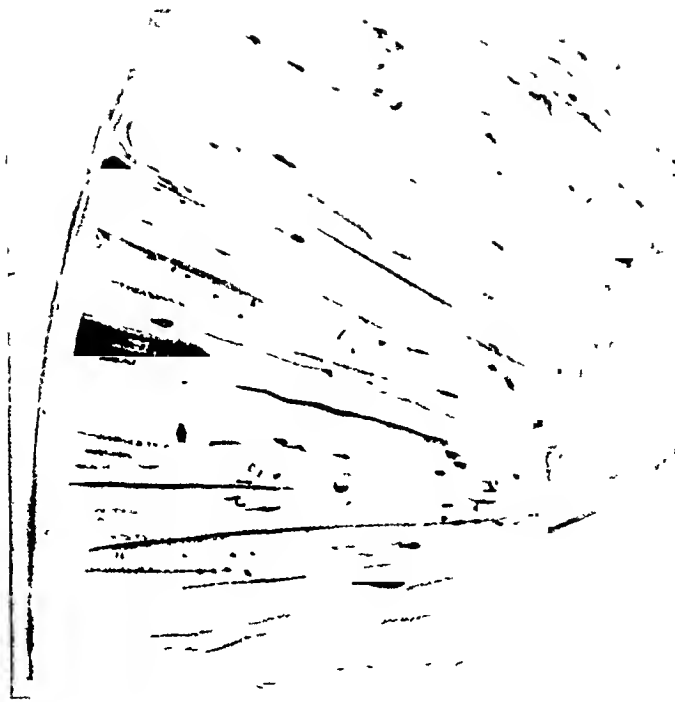


FIG. 462. Shrunken lens with stretched zonular fibers. Deposits on zonular fibers. Some of the fibers are missing.

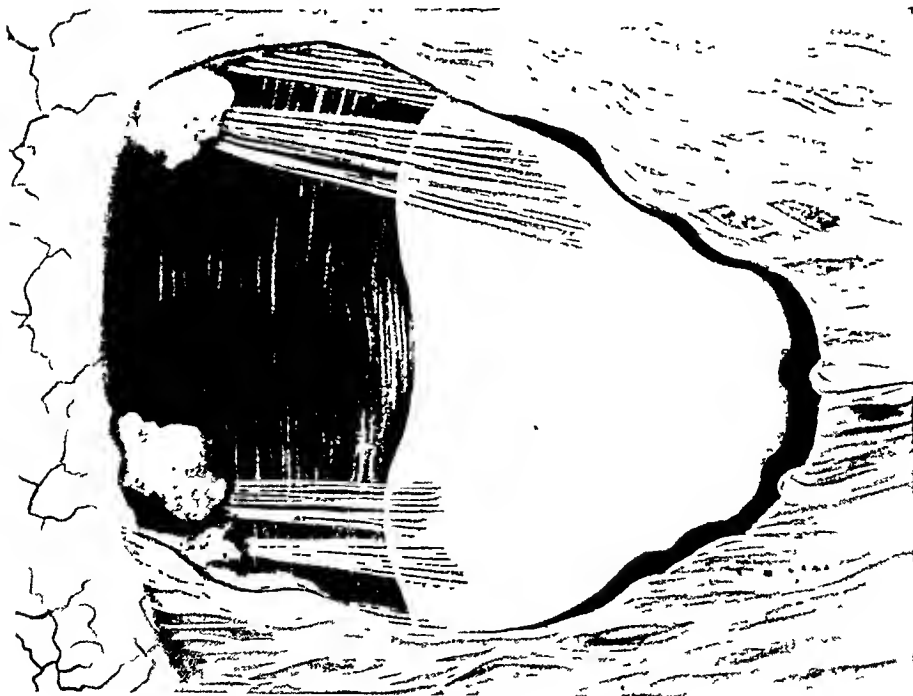


FIG. 463. Shrunken lens. (After Vogt.)

This striping has the character of zonular fibrillation. That their location is identical with that of the anterior hyaloid of the vitreous is demonstrated by the fact that these lines oscillate with it. They are seen in places devoid of zonular fibers and run either in the usual radial direction of the zonular fibers or at times concentric to the lens margin. The lines do not connect with the lens but seem to pass behind its edge. The nature and significance of these lines are not understood. Perhaps they represent adherent zonular fibers or possibly congenital rests. It is more difficult to explain those that run circularly or at right angles to the usual course of the zonular fibers excepting that they are reminiscent of the disputed occasional circular fibers connecting the ciliary processes (Eggers). In addition, particularly when the displaced or shrunken lens is opaque, its margin often casts a shadow on the surface of the vitreous adjacent to it. The width of this projected shadow increases as the angle of illumination is made more obtuse. Likewise the location of the shadow will wander with changes of the direction of the light.

TRAUMATIC ALTERATIONS OF THE ZONULE

The readiness with which the normal zonule tears is attested to not only in cataract operations (especially in older persons) but also by the frequency of subluxation and luxation even after moderate degrees of trauma. "The zonular attachment of the lens is of prime importance in maintaining the position of the lens. Much less important are the concavity of the fossa [action of negative pressure — M.L.B] and the connection of the lens to the vitreous by means of the ligamentum hyaloideocapsulare. If the zonule is ruptured completely or relaxed the lens shifts downward due to gravity but complete luxation does not take place until the ligamentum hyloideocapsulare is also torn" (Bellows).

As mentioned in the chapter on injuries to the lens, Frenkel believes that any sudden force applied to the globe which raises the intraocular pressure sufficiently will result in some degree of lens displacement (traumatic syndrome of the anterior segment). He postulates that minimal subluxations practically always occur in this

syndrome, although they may not be demonstrable in every case clinically. In this chapter we shall not consider those cases of severe injury or perforating corneal ulcer in which the lens is entirely



FIG. 464. Traumatic dislocation of the lens. Cortical changes (water slits and laminary separation). Note spheroid shape of the lens and absence of peripheral divergence resulting from the absence of zonular traction. Anterior surface of the vitreous seen above.

extruded from the eye, nor those in which, owing to rupture of the sclera, the lens is displaced into the extraocular tissues (in the subconjunctival space or within Tenon's capsule). Neither shall we dwell on those instances in which the lens is totally luxated into the vitreous and is not accessible to biomicroscopic examination. In certain types of injuries, associated with rupture or dialysis of the iris, operative coloboma (especially after excision of prolapsed iris tissue), shrinkage or lens displacement — it may be possible to inspect the zonular apparatus (Fig. 464).

The degree of zonular damage varies from case to case depending on the local conditions and on whether the lens is subluxated or luxated. In cases in which no discernible dislocation of the lens occurs, and an iris rupture or coloboma exists, the exposed part of the zonule may be studied *in situ*. The zonular bundles may be stretched, torn and curled or entirely absent. By changing the direction of the light (i.e., causing the light to enter the eye from the nasal as well as from the usual temporal side) it may be found that in places apparently devoid of fiber bundles, that the posterior ones are still

attached to the lens. Owing to tension these bundles are frequently wedge-shaped, i.e., the angle between them gets smaller the farther their distance from the lens margin. In addition to the deposits on or between the fibers (also seen in ectopia and spherophakia) additional ones may be present derived from the iris (traumatic iritis) or from blood cells (hemorrhage). In practically every contusion or perforating injury a positive tyndall phenomenon (pigment, white or red cells) in the anterior chamber will be present for a long time. Herniations of the vitreous are not uncommon.

When the lens is displaced, inequalities in depth of the anterior chamber will be noted, and especially in the young the edges of the lens may become undulated. (See Plate LXXVIII, figs. 2, 4, 5, 6.) In older persons this irregularity in contour of the lens equator is more apt to occur after the lens has become opacified. Iridodonesis and phakodonesis may be present. At times in lens displacement, when the tips of the ciliary processes can be seen in a coloboma or through peripheral iris defects, even when the remaining visible zonular fibers are taut, the tips of the ciliary processes may not be under any tension (Fig. 463). This is another indication disproving the older idea that the zonular fibers originated from the ciliary processes. Immediately after complete anterior luxation, if this is unobscured by hemorrhage, the lens released from its zonular attachments tends to appear more spherical than normal (Fig. 464). In diffuse light a clear lens may resemble an air bubble commonly seen after the insertion of a contact lens. However, the passage of the focal beam will reveal the pearly gray relucency normal to the lens (Fig. 460 A). Usually the iris will be displaced posteriorly. Occasionally an eye tolerates an anteriorly luxated lens quite well but in most instances, depending on the size of the lens and whether or not the aqueous circulation is interfered with, secondary glaucoma supervenes. Iridocyclitis either preceding, causing or accompanying the rise in intraocular pressure may further complicate the picture.*

* Several years ago, I saw a patient (a man aged 44) several weeks following a contusion in which the lens was luxated into the anterior chamber. The vision in this eye with a -4.00 D. lens was 20/50- and the intra-ocular pressure was 30 mm. Pilocarpine 2 per cent was instilled and the patient was referred to the hospital for operation. By the time the

Vogt describes an unusual phenomenon — separation of the capsule at the equator by fluid, occurring after a perforating injury to the eye. The opaque lens was subluxated into the anterior chamber. Below, stretched-out zonular bundles were seen to be attached to the lens margin. Some of the posterior bundles were present. With the optic section it was seen that the smoothly rounded capsule (with its zonular attachments) was separated from more opaque axial parts by a dark, less-relucent interval. Evidently in this case, owing to increased zonular tension, the total capsule itself was separated from the underlying equatorial lens substance by fluid. Vogt cites Topelansky who described, in postmortem specimens, collections of fluid at the equator that could occur only if the traction of the zonule were pathologically increased.

TRAUMATIC DETACHMENT OF THE ZONULAR LAMELLA

Traumatic detachment of the zonular lamella in the living eye was first described and seen biomicroscopically by Meesmann (1925). (See Fig 465; Plate LXXVIII, fig. 2.) The same year, Vogt described it in hereditary displacements of the lens and considered that there was a definite genetic predisposition for its occurrence (Fig. 466). In 1940 Serr confirmed Vogt's findings. Other writers, among whom were Stein, Busacca, Jess and Kronfeld, have described this condition. In Meesmann's cases (observed six months after injury) a "free floating crinkled membrane" was noted in the area of lens displacement, concentric to but separated from the lens margin. There were no zonular fibers visible. In Stein's case, in which there was a bilateral upward dislocation of the lens, concentric to the edge of the lens (but somewhat higher) there was a delicate pellicle suspended from the zonular fibers. Some of the fibers were seen freely floating in the anterior chamber. Jess described a case in which the detached

patient reached the operating table his pupil had dilated (psychogenic?) and the lens had fallen back into the vitreous. He was sent back to his room and instructed to lie on his face. The lens just as suddenly fell back into the anterior chamber and the pupil was vigorously contracted with a mixture of pilocarpine, eserine and doryl. With this treatment the lens remained in the anterior chamber and was extracted (with a loop) two days later. Although no vitreous escaped at the time of operation, biomicroscopic examination two weeks later revealed a large bulging herniation of the vitreous into the anterior chamber.



FIG. 465. Shrunken lens. Detached zonular lamella with adherent anterior zonular fibers.
 Shrunken opaque lens seen above. Zonular fibers are stretched.

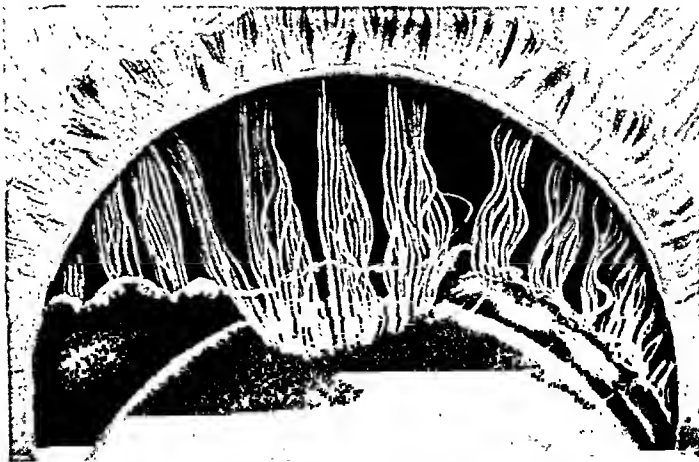


FIG. 466. Detachment of the zonular lamella. (After Vogt.)

zonular lamella and the attached zonular fibers were stained green by impregnation with copper (zonular chalcosis from an intra-ocular copper foreign body).

Undoubtedly more frequent biomicroscopic examinations in cases of displaced lenses either of hereditary, spontaneous, or traumatic origin will reveal that separation of the zonular lamella is not as rare as was first thought. As previously indicated, the individual nature of this lamella was proved by the fact that it can detach under stress, that this detachment is always limited to the equatorial zone, and that in trauma, unlike heat cataract * or senile cataract in which the detachment occurs axially, the zonular lamella does not separate in a flaky fashion.

* In heat cataract occasional flaking has been noted as a rare finding.

Chapter Thirty-One

THE VITREOUS

GENERAL CONSIDERATIONS

ALTHOUGH the fibrillar structure of the vitreous had previously been noted in aphakic eyes, it was not until 1911 that Gullstrand, using the focused beam, first noticed it in a normal living eye. He said that by oscillating the illuminating lens (which in the original model was held in the hand) it was possible to see several layers of membranes at various depths, and that these membranes formed a network and extended in the frontal plane. In 1914 Erggelet⁴²¹ studied the normal and pathologic vitreous in more detail. Later Redslob and especially Koby made definite contributions to biomicroscopy of the vitreous. In 1918 Vogt, employing a new method of projection (see Vol. I, page 5), had just demonstrated the principal of the optic section, obtained by means of a narrow beam. Thus it became possible to obtain sharp optic sections of the living cornea, lens, and vitreous, to the order of 15 μ . This advance became the cornerstone of biomicroscopy. In 1924, Bedell³⁶¹ elucidated the biomicroscopic appearance of the vitreous. His was the first important publication on this subject by an American writer.

Projection of the beam demonstrates a definite tyndall phenomenon in the vitreous. The fact that the tyndall phenomenon seen biomicroscopically is not homogeneous, as in the case of true colloids and gels, started a mild controversy concerning the physical structure of the vitreous. (See Vol. I, Figs. 6, 70.) Its lack of homogeneity is indicated by the presence of a framework consisting of multiple layers of vertically suspended bands or curtains, sometimes membranous and other times fibrillar, separated from one another by

darker intervals. The luminosity of the curtains gets progressively weaker in the depths. However, with stronger illuminants, e.g., arc-lamp, a fibrillar structure is seen even in the darker zones. It should be noted that the degree of luminosity varies in the normal eye from case to case. The more or less regularly arranged framework is best seen in the young.

Baurmann and Duke-Elder¹¹⁰ held that if the vitreous were a true gel, it would not be possible to discern any true fibrillar structure in the living state. They felt that since true gels are composed of ultramicroscopic micellae, the reflections which we do see, come from zones of varying indices of refraction and from a sort of *moiré*; the idea being that excepting by means of the *moiré* effect submicroscopic micellae could not be seen with the magnification afforded by the biomicroscope. The appearance of *moiré* (like watered silk) may result from reflections in which mirroring zones alternate with darker ones. A similar effect can be seen by reflection or transillumination in the case of parallel grilles or nets the elements of which cross one another angularly. According to Koby, in order for the vitreous to produce a *moiré* it should be formed of regular parallel elements—sufficiently opaque. This has not been proved. Even with the highest magnification possible and with the strong arc beam, no indication of a parallel crossing fiber system is apparent; such a system was described by Koeppe but has not been verified by any other investigator. Koby cites the fact that Duke-Elder,* following Baurmann, attributes great importance to the *moiré* (optical effect) which permits one to see ultramicroscopic fibrillae, but that Duke-Elder also points out that fibrillae are not visible in fresh

* After commenting on the fact that structures conforming to a fibrillar framework seen histologically can only be artefacts, or coagulation products resulting from fixation and staining processes, Duke-Elder adds, "The appearance seen with the slit-lamp requires a somewhat different explanation. In the beam of the slit-lamp the fibrillae only become evident when large numbers of them are arranged in a direction perpendicular to the incident light. In regions where the arrangement is haphazard, the vitreous appears optically empty, but near the boundary layers where the surface forces orient the micellae in approximately parallel directions and in closer formations, the optical effect is given of a waved or *moiré* appearance suggestive of watered silk or marcelled hair. The appearance of the vitreous is more than usually complicated, for as we shall see it contains internal surfaces within itself but the apparent texture is merely an optical effect, the basis of which is determined by the fibrillar micellae, although they themselves being of submicroscopic dimensions are too small to be rendered actually visible by the slit lamp."

specimens of vitreous. Thus Koby concludes that it is not possible to understand what produces the moiré and hence he believes that it plays no role in what is seen biomicroscopically in the vitreous. If the images were due to the effect of moiré then with movement of the globe or of the beam they should be displaced more rapidly than the anatomic elements. Actually this is not the case. With movements of the beam there is a definite lag and pendulum-like swing in the movements of the visible vitreous formations seen in the tyndall phenomenon. In my opinion the evidence seems to favor the conception that the formations seen in the tyndall beam biomicroscopically represent the true framework of the vitreous.

TECHNIQUE OF BIOMICROSCOPIC EXAMINATION OF THE VITREOUS

As pointed out by Koby, biomicroscopy of the vitreous is beset by difficulties not only attributable to optical inadequacies of the instrument but also because of the nature and location of vitreous itself. For example, its examination is hampered by the diaphragming action of the iris, its deep location and (chiefly owing to its gel-like character) its very weak relucency. Despite these limitations biomicroscopy is still the only available method for the direct examination of the vitreous in the living eye.

Because of the semifluid character of the vitreous, its examination biomicroscopically is restricted to observation by means of direct focal illumination. As in the case of the anterior chamber or even the zonule, morphologic details can only become visible by means of the tyndall phenomenon (Vol. I, page 73). Hence all observations in the vitreous have to be made within or along the passage of the focal beam. The low degree of relucency precludes the possibility of employing retro-illumination or indirect illumination excepting in light reflected from the fundus. Relatively the degree of relucency of the tyndall beam in the normal vitreous, excepting for its lack of homogeneity, is less than that of the normal cornea but is more comparable to that of a slightly turbid aqueous. The fundamental principle to be remembered when examining any part of the eye by direct focal illumination is that the focal part of

the beam and the focal point of the microscope must simultaneously be adjusted to fall on the spot observed. The gel-like structure of the vitreous, with its low degree of relucency, contains no regular stratification of zones of discontinuity, no regularly fixed points of reference. As the beam passes through the cornea and lens we are guided by well-defined surfaces and zones of discontinuity. These are absent in the vitreous. Its posterior surface is not visible normally and anteriorly there is no sharp line of demarcation — the posterior surface of the lens alone indicating its beginning. The focal point of the beam (convergent rays) where the light is most intense, depends on the focal length of the illuminating lens (70 and 100 mm.) augmented by the dioptric power of the media.* Hence it follows that using the ordinary biomicroscopic technique, i.e., without a contact lens, that only the anterior third of the vitreous can fall within the focal part of the beam. This limitation is not as serious as it may appear since most of the changes which occur are visible in these zones. Posteriorly the light (divergent rays) becomes weaker and is dispersed. However, further increase in depth of focus may be obtained by employing diaphragms of 70, 100 or 200 mm. (see Vol. I, page 48) now standard equipment in the Poser model (Bausch & Lomb).

Another handicap to deeper inspection of the vitreous lies in the fact that owing to limitations of the instrument we are unable to direct microscopic examination along the axis of the beam since the direction of the beam as it enters the eye is angulated to that of observation. This means that it is not possible to project the beam directly in the sagittal plane. To overcome this — to a degree — two maneuvers are resorted to: first, in order to cover as much area as possible, the beam is directed into the eye from the nasal side as well as the temporal, and second, the angle between observation and illumination can progressively be narrowed to almost 30 degrees. But as this angle is sharpened there comes a point at which the

* The chief advantage of the 100 mm. illuminating lens resides in that, although the intensity of illumination of the beam is somewhat lowered, its longer focus allows us to direct the beam across the nose without the end of the illuminating system touching the patient as might happen when using the 70 mm.

illuminating lens will strike the microscope and where it will obscure one of the objectives and thus interfere with binocular vision. In addition, as we narrow this angle specular reflexes from the cornea



FIG. 467. Fentoscopia. Appearance of the beam passing into the eye (observation with the unaided eye).

may cause annoyance. The binocular microscope commonly employed (Vol. I, page 28) has two paired objectives, the frontal ends of which converge so that there is a distance of 2 or 3 mm. between them. Consequently the distance between their optical centers measures from 12 to 13 mm. It will be readily realized that unless the pupil is widely dilated (at least to 5 mm.) the margins of the iris will prevent observation of the vitreous through one of the objectives i.e., the one farther away from the light. Koeppe, Lopez-Lacarere, Goldmann and Kleefeld have devised certain modifications to overcome some of these difficulties — but in each case added supplementary equipment (mirror or prism and contact lens) lowered the intensity of the focal beam. These modifications will be considered later. If they are not available one must not forget to use the ophthalmoscope with plus 4 to plus 9 lenses, since by this means opacities in the posterior part of the vitreous which are not over-delicate may be brought into view.

The beginner must also be cautioned about the possibility of striking the cornea when employing the longer high-powered objectives. To avoid this one adjusts the microscope so that the frontal ends of the objectives just clear the cornea and then safely focuses by racking the microscope backward. In view of all this, although a maximum magnification of $60\times$ can be used, practically a magnification of $24\times$ is sufficient to bring out even minimal pathologic changes.

I have always advised students to observe the passage of the beam with the unaided eye — fentoscopy (Koby) — since much information can be obtained by this oft-disregarded procedure (Fig. 467). Macroscopically it is possible to see gross disturbances in the vitreous structure. With eye movement, normal as well as pathologic trabecular formations are seen to oscillate. Cellular deposits give a definite increase in the tyndall dispersion, and even the color of the cellular components can be recognized. Actually the red color of blood can be recognized better macroscopically than microscopically. Under magnification, single erythrocytes appear as whitish or yellow-white, shining points. Another interesting feature that can be observed macroscopically is that while moving the beam across the pupil and directing the observer's vision along the axis of the beam, a reddish glow may be perceived which illuminates not only the beam (tyndall) but also the neighboring areas. This form of illumination (page 743 and page 981) results from light reflected from the fundus, which in a sense serves as a concave mirror. When detachment of the posterior limiting layer of the vitreous extends anteriorly, it can be diagnosed macroscopically.

Unquestionably the tyndall phenomenon, as obtained by the focused beam, is enhanced as the intensity of illumination is increased. Therefore early workers in this field resorted to the carbon-arc lamp as a light source. Although this does bring out considerably more details (especially when used in association with the narrow slit opening) practically, from the standpoint of everyday clinical work its use is not entirely without drawbacks. Even under the best circumstances the light is unsteady and requires constant adjustment. Stronger illumination can be obtained from the nitra lamp if it is overloaded up to 12 to 14 volts. However, I have found that with the stronger light source now available, i.e., with the Koeppe-Poser system, adequate illumination can be obtained. After focusing the posterior lens capsule — as a point of reference — using the narrow beam (optic section) the width of the slit should gradually be increased until details in the underlying vitreous become sufficiently visible, or conversely, one may focus on the posterior capsule

with the wide beam and then gradually narrow it until the posterior line of disjunction of the lens differentiates itself from the capsule stripe — an indication that an optic section of sufficiently small width has been reached. Also by overloading the lamp the tyndall phenomenon and hence the trabecular structure becomes increasingly visible. Even with the unaided eye it will be seen that the more luminous layers are interrupted by relatively dark ones. The anterior ones are most luminous and the most anterior is the most luminous of all. The presence of such variations in the tyndall dispersion of the vitreous proves that it is not homogeneous, as would be expected if it were a true gel. Not infrequently, particularly in cases where the vitreous (even when it is normal) is of low relucency the slit may have to be opened maximally. The conical bundle as obtained from the round openings in the diaphragm is also useful (as in the anterior chamber) for finding varying degrees of relucency (at different levels) and for localization. Koby recommends the astigmatic bundle (see Vol. I, page 21) because of its homogeneity and increased luminosity (three times that of the ordinary one obtained with Vogt's method of projection). However, the height of the beam becomes disturbingly elongated.

As pointed out in the section on technique of biomicroscopy (Vol. I, page 64) it is not necessary to keep the examining room in total darkness — sufficient light should be present to permit the examiner to make adjustments in the apparatus and to watch that the patient keeps his head steady by means of both the chin rest and the head guard. It is preferable to have the patient gaze directly ahead rather than toward the microscope or into the light. Also it is better to make changes in directions of illumination and observation by altering the relative position of the microscope and illuminating lens, than to have him constantly change the direction of his gaze — since ordinarily on command his excursion will be too wide. However, there are times when slight movements (especially vertical) will serve to bring out certain details (see page 1379). Without employing the microscope, the beam of medium width is projected into the dilated pupil (when dilatation is feasible) — first from the

temporal side and then from the nasal side; the beam is then brought to a sharp focus at the posterior capsule. The tyndall effect of the beam is then studied as the intensity of the light is increased, i.e., by widening the slit and by decreasing the resistance of the rheostat. By looking somewhat over or below the beam, the observer with the unaided eye can direct his gaze along the axis of the beam, a procedure not easily possible when using the microscope. The beginner will frequently be surprised by the fact that a tyndall effect in the vitreous may be obtained even in the presence of considerable change in the lens — although dense central opacification will tend to impede it more than peripheral ones. Corneal opacification offers greater hindrance. After having made observations by means of the naked eye, the examiner proceeds to observe the tyndall by means of the microscope — starting with low powers of magnification. In order to obtain deep penetration the angle between illumination and observation should be as narrow as possible. Again, changes in direction and intensity of the beam should be made in order to cover as much area as possible and in order to bring out the maximum tyndall effect as strongly as possible. As the direction of beam is slightly displaced from side to side, one will notice that the more luminous constituents (curtains) move naturally in the same direction but not exactly at the same rate of speed. When pathologic changes are found, it is always good practice to study the fellow eye immediately — for the purposes of comparison. Quick changes in observation from eye to eye will serve to accent the relative differences in appearance. As Bedell (1924) has stressed, prolonged and leisurely observation of the vitreous very frequently brings out parts that otherwise would be glossed over in more hurried inspections.

BIOMICROSCOPIC APPEARANCE OF THE NORMAL VITREOUS

Normally the structure and luminosity of the vitreous is extremely variable even in persons of the same age. Its appearance also changes as the individual grows older, and eventually in the aged it may assume characteristics which by themselves may not be differentiated from changes due to pathologic processes.) Variability in the

relucency of the vitreous tyndall phenomena may be so great that extremes in the degree of visibility of the framework result. In one case the latter may be practically invisible, while in another it may



FIG. 468. Diagrammatic representation of the normal vitreous. To the left, posterior lens capsule with the attached remains of the hyaloid vessel. The latter is seen suspended in the retrolental space or in the hyaloid vitreous (*H.V.*). The hyaloid (primary) vitreous is composed of delicate striae. Deeper is the definitive (secondary) vitreous (*D.V.*) with its plicated surface (*membrana plicata*) well demarcated.

be highly luminous and easily seen. Such contrasts in normal eyes probably result from varying differences in the indices of refraction between the component parts of the vitreous. When the difference in indices of refraction between the framework and its more fluid parts is minute, then the visibility will be very low. For purposes of illustration Koby cites the work of Veragut, who, with the nitra lamp, roughly estimated variations in relucency of the vitreous in a group of 82 children as follows: very luminous framework, 6 per cent; fairly luminous framework, 44 per cent; slightly luminous framework, 50 per cent.

As the beam penetrates the vitreous, an optic section (tyndall phenomenon) is formed. The width and relative luminosity of this section depend on the width of the slit opening and intensity of the

beam. With the nitra lamp, a dark space (so-called "retrolental space") lies between it and the posterior capsule. However, with the arc lamp (Erggelet) or the stronger illumination now available



FIG. 469. Frontal view of membrana plicata showing details under high power.

with the Poser apparatus, it will be found that a faint tyndall effect occurs in this dark area and demonstrates a fine fibrillar network — in other words vitreous and not simple aqueous (Fig. 468). This may represent the embryonal primary vitreous (hyaloid) — an idea strengthened by the frequent finding of traversing vascular remains. Immediately behind this area is the definitive or main part of the vitreous (secondary) which is indicated by the appearance of a more definite tyndall effect and the plicated membrane.

(Immediately behind the so-called "retrolental space," luminous "layers" or "bands" appear (in lieu of a better name for something uncertain) which run more or less parallel and are interrupted by narrower, less relucant or dark layers. These so-called "dark" spaces are only so relatively; with higher degrees of illumination and with

changes in direction of the beam they are not optically homogeneous but contain delicate fibrillar structures. The transition from a dark to lighter area occurs rapidly in a "sinuous line" upon movements of the beam. The anterior bands are more luminous and regular than those behind them which gradually fade in the depths, the most anterior having the highest luminosity (Fig. 469). These curtain-like bands hang vertically, their direction having no relationship to the direction of light. Changes in position of the head or eye will cause them to float but, owing to gravity, they will always resume their vertically hanging position.)

According to Koby, variations in the appearance of the anterior band make it possible to distinguish two extreme types of structure. Thus two schools of thought arose. This apparently resulted from variation in density of the luminous parts. The first group (Koeppé, Koby, and Mann) postulated that the vitreous had a fibrillar structure and the second (Gullstrand, Erggelet, Vogt, Dejean) that it was lamellar, pseudo lamellar or membranous, later Koby stated that both forms occur (Figs. 470, 471, 472). Both groups agree that the luminous parts are reflections from zones of discontinuity. But as Koby asks, "To what do the darker or less luminous parts correspond?" Those that consider the structure to be membranous think that the dark parts are due to unlighted portions of the folds. In this case, lateral movements of the light should cause the dark parts to become luminous. It has not been possible to demonstrate this. Koby thinks that the alternating light and dark areas may represent uniform differences of density in the same surface or perhaps, as suggested by Redslob, because of an irregular arrangement in layers, certain areas become more dense and hence more luminous. In this way the alternating dark and light parts could be explained partly by the presence of true folds in the pseudomembrane and partly by the heterogeneity of its structure. As Koby puts it, in spite of the several theories concerning the structure of the vitreous — i.e., whether it is formed of homogeneous membranes, isolated fibers, or other unique colloidal structures — not one of them explains in an entirely satisfactory way its biomicroscopic ap-



FIG. 470. Normal vitreous of membranous type. Young person. Lens normal. Remnants of hyaloid vessels attached to the posterior lens capsule. Main remnant hanging dependently in the hyaloid vitreous, which is characterized by a fine oblique striation. The anterior band or surface of the definitive vitreous is very luminous, appearing as a finely pleated membrane (*membrana plicata*). Deeper there is another pseudomembrane, less luminous and less well defined. (After Koby.)

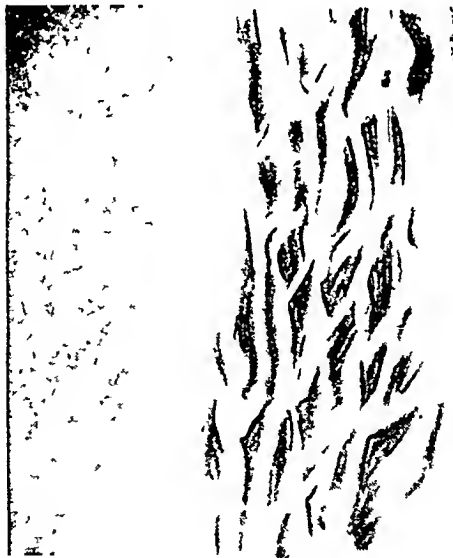


FIG. 471. Normal vitreous of the membranous type.

pearance and physical properties. It is the biomicroscope which provides the most realistic picture and no theory concerning its structure is acceptable unless it harmonizes with this picture.



FIG. 472. Normal vitreous of the fibrillar type. Young person. Notice the numerous obliquely running bands which are finely striated and seem to anastomose in some places.

Vogt has compared the frontal reflecting bands of the vitreous framework to hanging drapes, vertically folded. The first of the anterior bands, being the most luminous, shows the angulated folds best. This is sometimes known as the plicated membrane (Fig. 469). It probably represents the anterior boundary of the secondary vitreous and hence there is good reason to believe that it corresponds to the upper boundary of Cloquet's canal. In agreement with Fuchs, (Vogt considers that the vitreous has a lamellar or membranous structure and that the lamellae in turn are composed of fibrillae. The fibrillar structure becomes especially apparent in pathologic states. Vogt intimates that a purely fibrillar structure may be seen in many normal eyes but that with increased illumination their membranous character becomes more apparent. Evidently the form and position of these scaffolding membranes or perhaps better pseudo-membranes, is a result of their specific weight, which is greater than that of the surrounding vitreous fluid. Owing to some degree of fixation since the scaffold is probably attached to and suspended

from the so-called base of the vitreous (Salzmann) in the region of the flat portion of the ciliary body, movements of the eye cause it to have pendulum-like movements. But in these movements it quickly returns to its original position. In addition to the vertical folds Vogt called attention to crossing stripes (due to horizontal folds?) which gives the normal framework its characteristic appearance.) Koeppé, using the Nernst-lamp, described systems of regular nets consisting of parallel crossed fibers (horizontal and vertical). No other investigators have been able to verify the presence of such regular systems. As the deeper parts of the vitreous are brought into focus the vitreous bands become less luminous and less orderly. Frequently it will be observed that the vertical and parallel arrangement of these structures is limited by the first two or three bands. (The deeper ones become irregularly tortuous and sinuous so that no regular directions can be attributed to them.) Consequently they lose the curtain-like effect of the frontal layers so characteristic of the normal vitreous structure in the young.) With slight movements of the beam, this irregularity gives the impression that connections exist between the lamellae and that perhaps in these parts no orderly stratification exists. As the light is angulated, it will be seen that peripherally (as far as is possible to see) the relucency of the tyndall effect may be greater than in the axial region, the structure appearing more fibrillar. (From the standpoint of biomicroscopic appearance it may be said that the vitreous is a complex gel-like substance, having a structure or framework in the form of loose, nonhomogeneous layers, lamellae, or pseudomembranes; that these lamellae, which vary in thickness, are constituted of irregular conglomerations of fibrillae; that in it we can see vascular rests and divisions (i.e., the retrolental space with its contained hyaloid or primary vitreous and the definitive or secondary vitreous) indicative of its embryologic development.)

THE RETROLENTAL SPACE AND THE HYALOID VITREOUS

The so-called "retrolental space" is bounded anteriorly by the posterior surface of the lens and posteriorly by the first band of the secondary, or definitive, vitreous. (See Figs. 468, 470, 472.) In cer-

tain pathologic conditions the division between these parts becomes uncertain. This space can easily be seen even with the unaided eye. Focus of the beam upon the posterior lens capsule will reveal a dark interval between the back of the lens and the plane marking the onset of the vitreous tyndall effect. This retrolental area also is known as the "space of Berger." Depending on the posterior curvature of the lens, it will be seen that this space is shallower axially and gradually increases in depth toward the periphery. Its extreme peripheral boundaries cannot be seen. Some of the earlier writers considered the retrolental space to be "optically empty," its relucency corresponding to that of the aqueous. As previously mentioned, with improved illumination (especially with the arc lamp) it soon became apparent that this area contained a delicate fibrillar striation.* Even with the ordinary nitra lamp, if the observer is slightly dark adapted and persistent (similar to the conditions required for bringing out the physiologic flare of the anterior chamber), he will, after gradually increasing the intensity of illumination, suddenly become aware of this delicate fibrillar striation. These fibers in traversing the space tend to run obliquely from above downward (Fig. 472). According to Koby, the contents of this area are not generally homogenous, and frequently two or three pockets of varying optical densities will be found. It should be emphasized that at best the tyndall effect in the retrolental space (primary or hyaloid vitreous) falls just within the limits of visibility and hence does not interfere with the sharp demarcation of the less homogeneous secondary or definitive vitreous behind it.

During embryonic development the primary vitreous contains the hyaloid system of vessels. Later, as a consequence of either pressure of the secondary vitreous or condensation of its fibrillae, the boundaries of Cloquet's canal are formed. In this way the primary vitreous becomes enclosed. According to Ida Mann the direction of the vestiges of the vessels and the boundaries of Cloquet's canal in the fetus and even for some time later in the young have a some-

* This was first emphasized by Erggelet. It will be readily understood that the depth and dark contrast of this space becomes greater with less illumination.

what sagittal direction. With time, the upper boundary of Cloquet's canal becomes increasingly convex frontally and with its vascular remains sinks so that gradually a vertical direction is assumed (Fig.

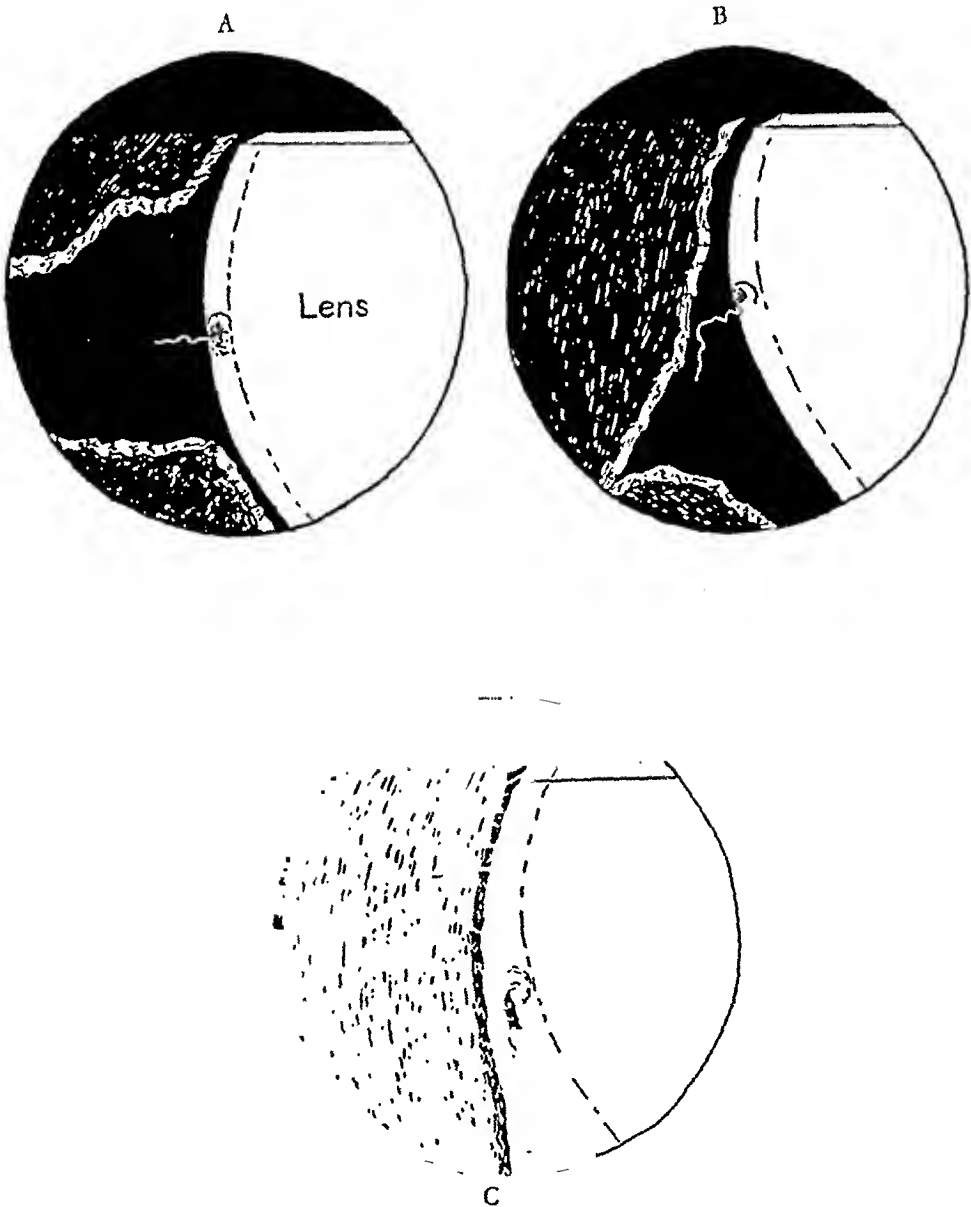


FIG. 473. A. Cloquet's canal in the newborn. The "entenoir" of Cloquet's canal is very large, the hyaloid vestige is horizontal in direction. B. Cloquet's canal in a young person. C. The effacement of the canal in an adult. (After Mann.)

473). In other words, the vertical anterior band (originally the upper boundary of Cloquet's canal) now limits posteriorly the so-called "retrolental space" with its contained vestige of the primary vitreous and vasa hyaloidea propria. It is therefore appropriate to

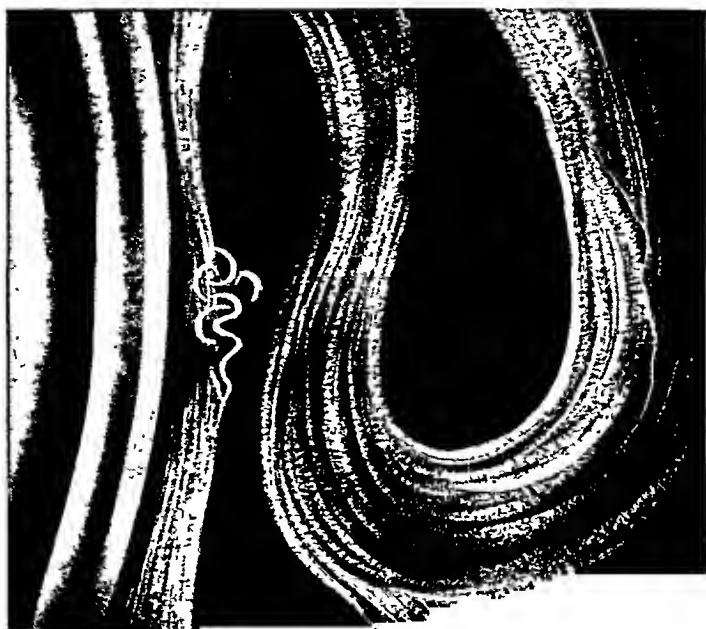


FIG. 474. Dark pocket above Cloquet's canal in myopia. Opacity in shape of a veil in the case of retinal detachment. Behind posterior capsule of the lens there is a zone of increased optical reluctance formed of delicate fibrils. Deeper to this there is a darker zone in which remnants of the hyaloid vessels are seen. Still deeper there is a thick layer seemingly formed by a great number of fine membranes. This layer then turns upwards enclosing a cul de sac—optically empty. The limiting posterior layer of the pocket appears to be especially luminous and fibrous. (Modified after Koby.)

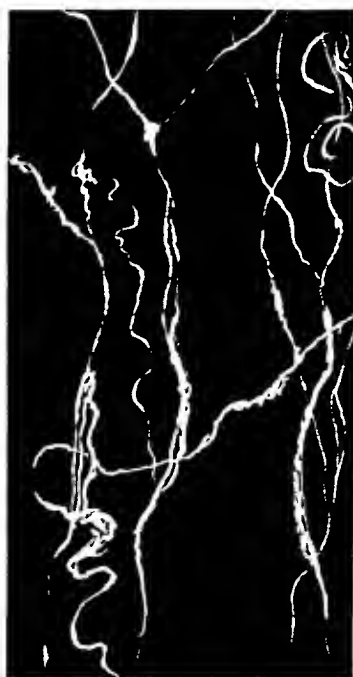


FIG. 475. Rests of the hyaloid vessels located deep in the hyaloid vitreous. Note condensations where the fibers cross or anastomose.

call this the "hyaloid vitreous." At the same time it forms the anterior limit of the more relucient deeper secondary or definitive vitreous. The above phenomenon also explains the observation made

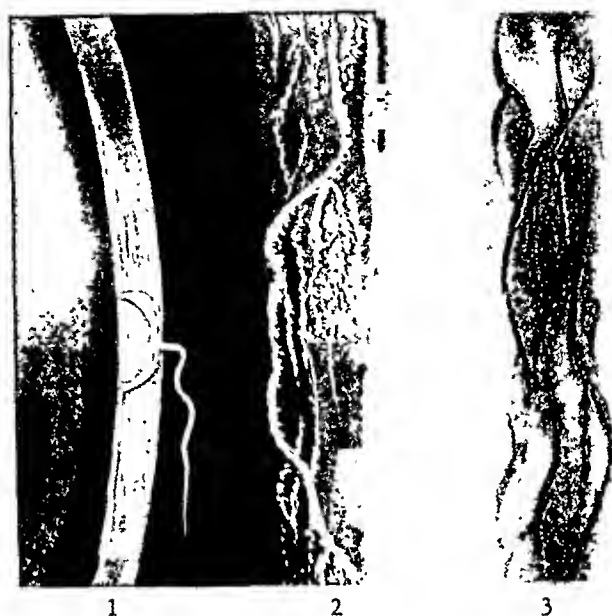


FIG. 476. Filiform rests of the hyaloid vessels located on the surface of the definitive vitreous and confined to the frontal plane. The retrolental space 1 is dark, deeper to this there is a wider zone 2 which contains wormlike filiform and anastomosing opacities. Still deeper (3) is another zone of lesser relucency with linear figures.

by Koby, that the direction of the anterior band of the definitive vitreous is somewhat obliquely backward from above down and thus causes the depth of the retrolental space to be greater below than above (Fig. 472). By having the patient direct his gaze upward and then back to the horizontal level, it frequently is possible for an instant to cause the anterior vertical band to float back to the original embryonic (?) horizontal direction. By focusing below this, another wavy horizontally directed band may appear, which, perhaps, represents the inferior limit of the canal. Koby has also described the finding of invaginated pockets, especially in myopes. These pockets, located in the anterior parts of the definitive vitreous, appear almost optically empty. They resemble the letter U, with the opening directed superiorly (Fig. 474). He believes that they are related to Cloquet's canal. Such appearances might also be associated with detachments of vitreous (page 1410). Another fact

strengthening the conception that the vitreous in the retrolental space corresponds to the embryologic primary vitreous is the almost universal finding of irregular filiform structures that are not attached to the lens. They may lie on the surface of the anterior band (*membrana plicata*). (See Figs. 475, 476.) They tend to run more or less vertically and frequently are "somewhat entangled and anastomosed." It is generally agreed that these threadlike structures are vestiges of the *vasa hyaloidea propria*. They are easily distinguished from the framework of the definitive vitreous by their more intense white color and ravelled thready character. Upon vertical eye movements these as well as the small corkscrew filament which is so often found attached to the posterior lens surface can be caused to float out horizontally. The latter returns only very slowly to its dependent position. The remains of the vascular system which are found attached to the posterior capsule were discussed in the section on the lens. In the normal eye, I have never seen any portion of the definitive vitreous attached to the lens. However, this may occur when the vitreous is altered pathologically.

SPECIAL METHODS DEvised FOR THE EXAMINATION OF THE VITREOUS AND FUNDUS

In order to observe the posterior two-thirds of the vitreous, Koeppe applied the principles of "orthoscopy" in the form of a contact lens of about -54 D. As a result, the convergent action of the anterior media (cornea and lens) was counteracted in such a way that, optically, the posterior parts of the eye were brought within the field of observation (Fig. 477). To avoid the pupillary border of the iris, the beam of light had to be directed deeply into the eye, thus forming a very acute angle with the anteroposterior diameter of the eye. This was accomplished by a tiny mirror affixed to the distal extremity of the arm of the slit lamp, the latter being directed more or less tangentially to the observed cornea. However, with this method, Koeppe was unable to employ the strong objectives of the ordinary Czapski binocular microscope. This was due to the fact that the mirror which was attached to the illuminat-

ing lens either completely or partially covered one of the objectives. Hence binocular vision, so vital to this method, was interfered with. Therefore he decided to replace the Czapski by the

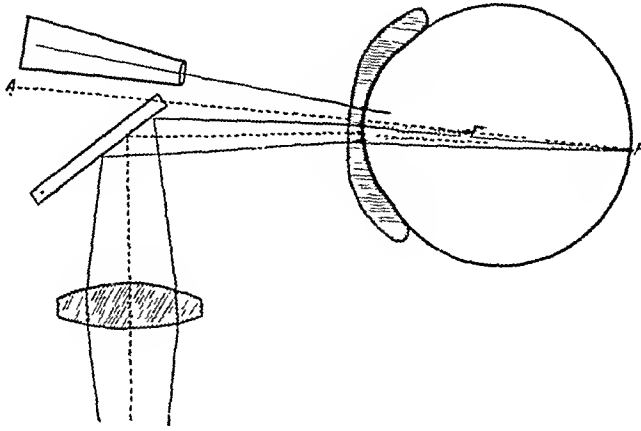


FIG. 477. Diagram of Koeppé's method for the examination of the retina and deeper vitreous.

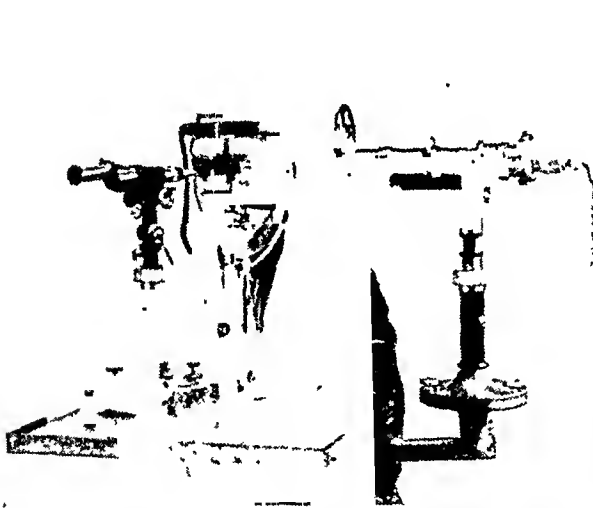


FIG. 478

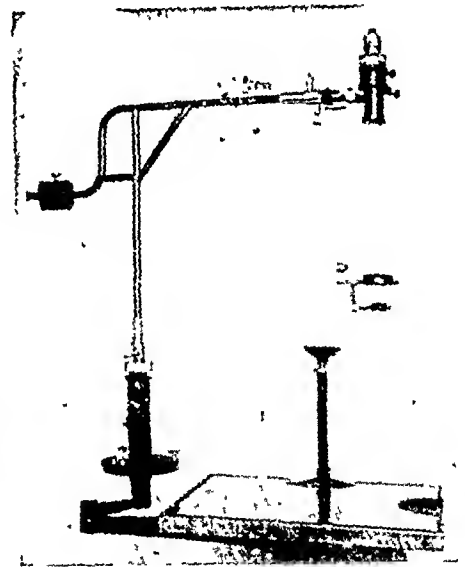


FIG. 479

FIG. 478. Koeppé's method—instrument ready for use.

FIG. 479. Kleefeld's instrument. A modification of the Koeppé method.

uniobjective binocular microscope built by Seidentopf and called Bitumi (Zeiss). (See Vol. I, page 28.) With this microscope the light enters the objective and then is split by prisms so that half is received by each ocular. All Koeppé's studies with respect to the vitreous body, and the fundus oculi as well, were done by means of this method (Fig. 478).

It should be remembered that Wolff (1912) (see Vol. I, page 5) was the first to employ a method for examination of the fundus by means of focal light. Other types of ophthalmoscope permitting the

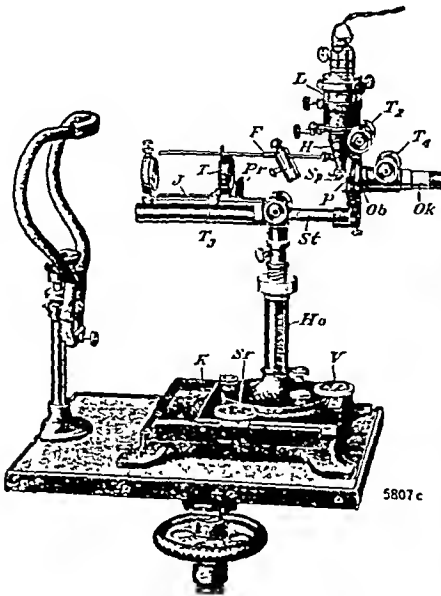


FIG. 480

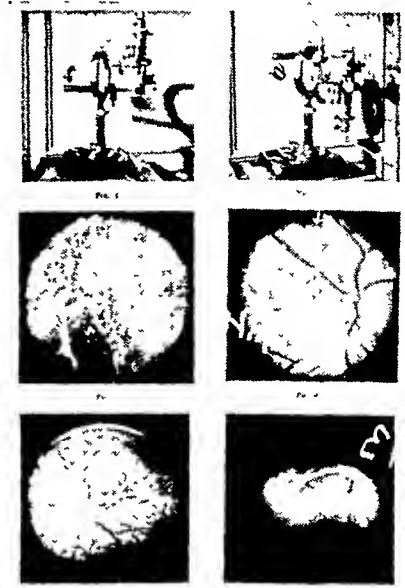


FIG. 481

FIG. 480. Zamenhof's modification of the simplified Gullstrand ophthalmoscope.
FIG. 481. Above: Kleefeld's modification of Zamenhof's method. Below: Photographs of different vitreous opacities. (After Kleefeld.)

use of a focal slit beam were developed by Thorner, Friedenwald and Evans.

Shortly after the publication of Koeppe's paper, Kleefeld (1922), changing the direction of the beam, proved that the Czapski microscope could be used with a contact lens. By having the light source above and by directing the beam horizontally into the eye by means of a mirror, it was possible to get an unobstructed view of the deeper vitreous. The mirror was attached to the end of the slit lamp arm and not to the illuminating lens (Fig. 479) as is the case in the Koeppe method.

The contact lens enables the use of high magnification. Without unnecessarily increasing the strength of the oculars a magnification of 36 diameters is obtained easily with good stereoscopic vision.

In 1935, Lopez Lacarrère had an apparatus constructed in which both the illuminating and observational systems were placed on a single supporting arm. This permitted observation along the beam's

axis. Also, with this objective in mind Lopez Enriquez (1935) employed two plane silvered mirrors.

Recently, Lindner (1936) devised a simplified monocular microscope having a reflecting prism which permits, to a degree, observation along the axis of the beam. A contact lens is also used. I have tried both the Koeppe and Lindner methods and must confess that I agree with other workers in this field that they are difficult and uncertain. For one thing the application of the contact lens tends to contract the pupil. One also must constantly guard against fogging of the contact lens itself, and with the patient's head in the upright position it is necessary to support the glass in order to keep it from slipping. In addition disturbing specular reflexes from the glass interfere with observations.* With the Lindner microscope I was unable to obtain sufficient magnification.

Zamenhof, adding a movable lens (+ 32 diophers) to the Simplified Gullstrand ophthalmoscope, devised a method for examining the vitreous, and the fundus as well, without a contact lens (Fig. 480). By the addition of the lens, he was able to obtain a focal beam. The binocular stereoscope system of this instrument gives a magnification of $24\times$. Although this magnification should be sufficient, fewer details are observed than with the contact glass methods. Nevertheless it is possible to study, for instance, detachment of the vitreous. With the Zamenhof modification of the simple Gullstrand ophthalmoscope it is difficult to examine the lower parts of the vitreous body because of the dark shadow in the lower part of the pupil. With the aid of a metal ring Kleefeld, in 1937,⁴⁹⁶ modified this apparatus so that it was possible to inspect the lower part of the vitreous as well (Fig. 481). In addition, he suggested a method for localization, mensuration and even photography of vitreous opacities. It should be noted that in all these methods in which the illumination penetrates to the posterior part of the eye, the pink

* Modern molded contact lenses fitted beneath the eyelids may obviate this objection to a certain degree. The use of wetting agents offsets the fogging of the contact lens which hitherto has been a considerable difficulty. Furthermore the patient may be examined in a sitting position. Recently Bausch and Lomb have constructed a plastic contact lens similar in principal to the one devised by Allen for gonioscopy. At the time of writing this was in the experimental stage.

reflex of the fundus, as in ophthalmoscopy, interferes with observations of delicate vitreous changes. The latter can be seen only by virtue of diffraction circles, an optical phenomenon dependent on

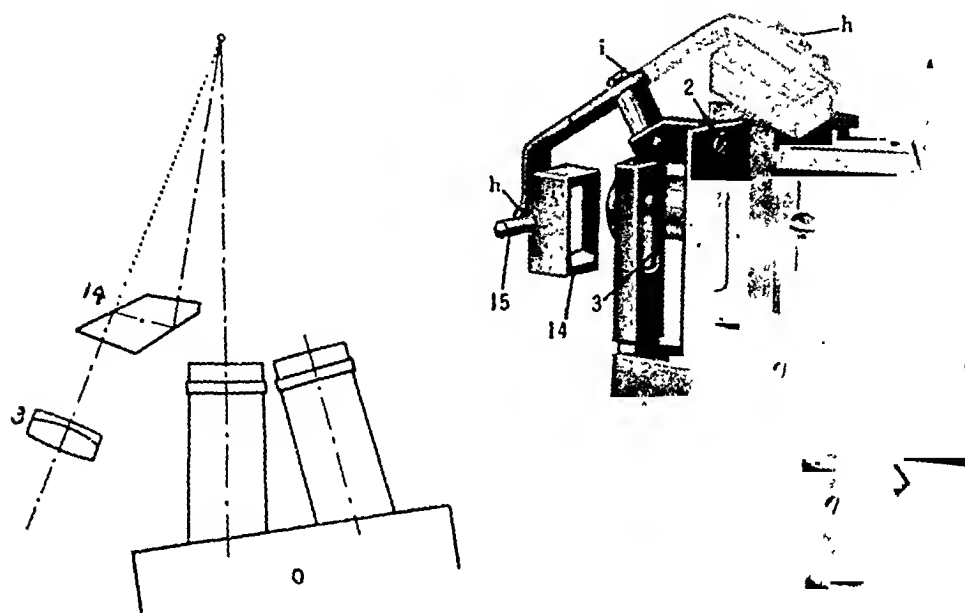


FIG. 482. A. Goldman angle reduction prism: 3, Illuminating lens at end of slit lamp arm; 14, four-sided prism showing double internal reflection giving reduction of the angle between the illuminating beam and axis of the microscope. B. The angle reduction prism attached to the slit lamp; 2, rectangular diaphragm on arm of illuminating system; 3, illuminating lens; 15, lever for rotating prism through 180 degrees; *i*, central mounting pivot allowing prism to be swung into alternative sections *h* and *h'*, whether in use or not.

the tyndall effect and requiring a focused beam against a dark background.

In 1937, Goldman demonstrated for the first time at the International Ophthalmological Congress in Cairo an improved biomicroscope produced by the firm of Haag-Streit. (See Vol. I., page 56.) By means of a prism and a contact lens, examination of posterior sections of the vitreous and fundus in focal light became feasible and practical. For the purpose of satisfactorily reducing the angle between the light beam and the direction of observation (which must be very acute) he employed a four-sided prism placed in front of the illuminating lens. The arrangement is illustrated in Fig. 482 A. The light beam, on leaving the illuminating lens, undergoes double internal reflection in this prism and is so diverted that the effective angle between the axis of illumination and that of the microscope can be reduced by about 13° . The prism (14) is rotatable through

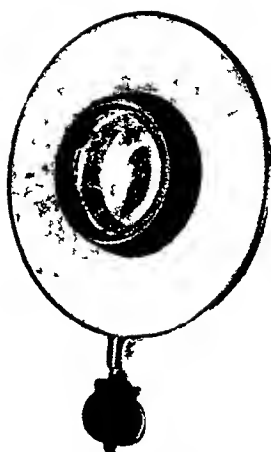
180° around the principal axis of the illuminating beam as shown in Figure 482 B by means of the lever (15). With the prism the intensity of the diverted light beam is greater than that obtained with a mirror.

In conjunction with this system Goldmann uses a contact lens made of artificial glass (plexiglass). This lens is very light in weight (0.8 grams) and hence is easily retained with the patient's head in the upright position. Recently he designed such a contact lens having two depressions located in the horizontal meridian so as to facilitate easy insertion and removal. This contact lens has a diameter of 20 mm. with an anterior worked surface 12 mm. wide, having a radius of 70 mm.

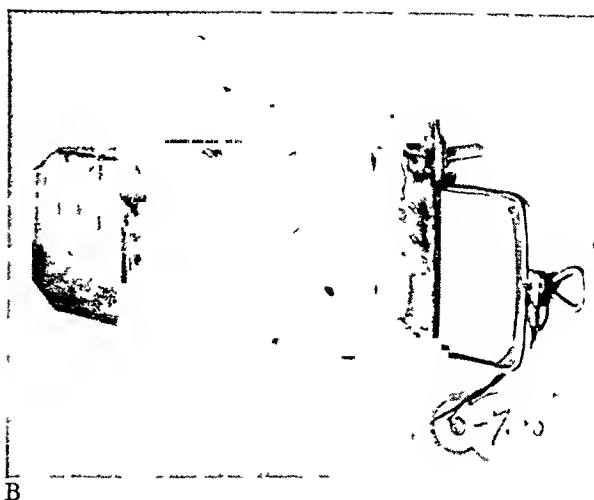
It is quite possible to obtain a satisfactory view (in focal illumination) of the deeper vitreous and fundus without the use of a contact lens. In 1923 Lemoine and Valois suggested this possibility. They placed a strong concave lens in front of the patient's eye. However, their system of illumination was diffuse. Lately Hruby (Figs. 483) using a focal beam, employed such a lens (—54 diopters with its anterior surface +4 diopters) to lessen reflection. I have employed such lens mounted in a trial frame and placed directly in front of the patient's eye. The axis of the beam was deviated by means of Koeppé mirrors (standard equipment of the Poser slit lamp, Bausch & Lomb; see Vol. I, page 50 and Fig. 48) or by a modified prism (still experimental, Berliner-Priestley).

The ability to obtain optic sections (narrow beam) in the examination of the posterior parts of the vitreous and retina will certainly assist us in clearing up many hitherto questionable points in diagnosis as well as elucidating the mechanism with which such changes develop. Although the use of the Hruby lens obviates the need for a contact lens, on the other hand the size of the field of vision obtained by means of it is smaller than that afforded by the contact lens. Also it is more difficult to view the myopic fundus with the Hruby lens.

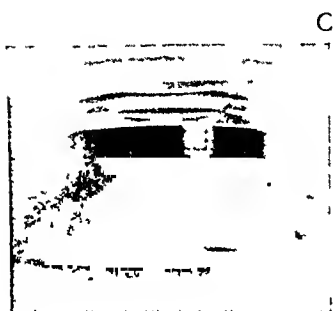
For example, it is possible to localize the posterior limiting layer of the vitreous; to see changes in retinal thickness, to determine elevations or depressions in the retina (particularly in the fovea)



A



B



C

FIG. 483 A. Hruby diagnostic lens (courtesy of A. H. Parsons Laboratories, San Francisco) B Berliner-Priestley prism. C. Contact lens (Goldmann).

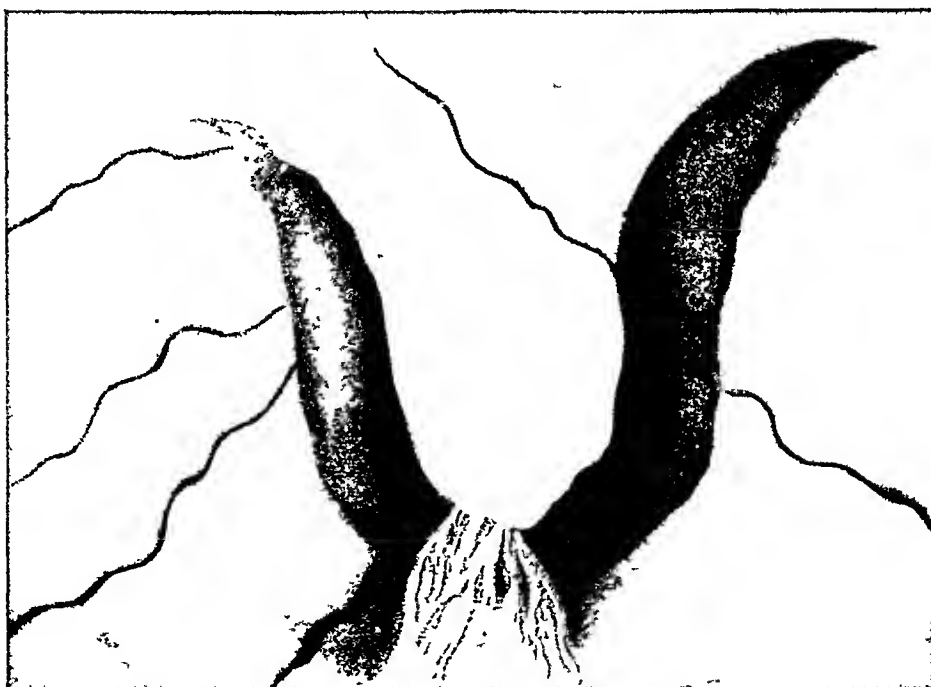


FIG. 484. Retinal tear as seen with the Hruby glass. Note strand of vitreous below, pulling tongue of retina forward. Retina detached.

and especially to diagnose separation or adhesions between the vitreous and retina, and similar alterations between retina and choroid. In a case of retinal detachment (Fig. 484) I was able to observe a tag of vitreous adherent to and exerting forward traction on the tongue of a retinal tear.

Through the courtesy of Prof. Dr. Hans Goldmann I am reproducing several interesting cases from his clinic reported on by Katharina Baumgarten-Blaser (Plate LXXIX, figs. 1-6). In this report the pictures seen by means of ordinary and red-free ophthalmoscopy were compared with those seen by means of focal illumination (narrow beam) and contact lens. In all instances certain important details could only be recognized by the latter method. The observations were made with the Goldmann apparatus and contact lens.

CONGENITAL ANOMALIES

Congenital anomalies of the vitreous, as far as they can be recognized, are all related to vestiges of the fetal vascular system (hyaloid arteries and its tributaries). The filiform threads behind the lens, the arcuate line, the hyaloid corpuscle, and the attached corkscrew-like remains of the hyaloid artery on the posterior lens capsule are so universal that they can be considered as physiologic. These have been discussed in the section on the lens (page 1024). Hence, in this section only those vestiges that have unusual forms or location will be considered. As suggested by Reese and Payne (page 1394) all these anomalies probably represent forms of abnormalities of the primary vitreous. Examination of the literature reveals only a few reports concerning such anomalous remains. Three main types will be described: (1) striate and pigmented stars on the posterior capsule, (2) the so-called retrolental fibroplasia and (3) congenital vitreous cysts (free-floating or fixed).

Striate or Linear Figures. These may be seen either localized or disseminated on the posterior lens capsule in the form of regular striae or curved lines, grayish white in color. When closely matted, they may form star figures or larger netlike membranes. Like the

PLATE LXXIX*

FIG. 1. Retinal detachment with macular cyst, three years following injury. Flat retinal detachment extending from above to below disc and macula (retinal folds). Macula region raised and surrounded by area of small honeycomb cysts. Question as to the presence of a hole or elevation in macula.

Slit-lamp: With the optic section the protrusion in the macula can be seen as an elevated line. Many small cysts are visible. The superficial line of the light beam is not interrupted over the central area but rather becomes threadlike and more convex. This would indicate the presence of a large cyst with an extremely thin anterior wall. In the region of the cyst the beam can be clearly seen while adjacently it becomes more hazy. More peripherally the beam becomes sharper again indicating a flat detachment. In the central cyst the choroidal line has a yellowish tint which suggests that it is in the region of the macula lutea and that it includes some retinal tissue. Small white spot on the anterior wall cyst is evidently caused by a thickening of tissue about the end of the vessel.

FIG. 2. Senile macular degeneration with hole formation. Male 76 years of age. Thrombosis of branch of lower temporal vein (not seen in illustration).

Slit-lamp: The narrow beam (light directed from the left) shows a round defect comprising all the retinal layers. The choroidal line of illumination (more yellow) proceeds without interruption across the bottom of the hole indicating that the floor is formed of choroid. The floor is tigroid in character.

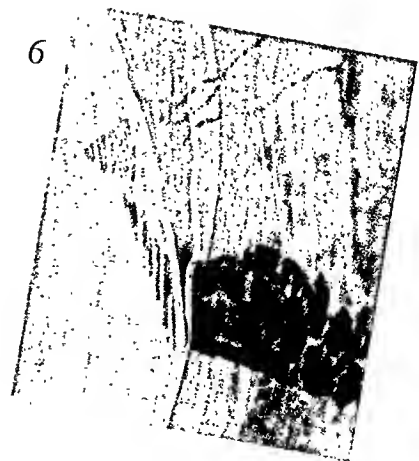
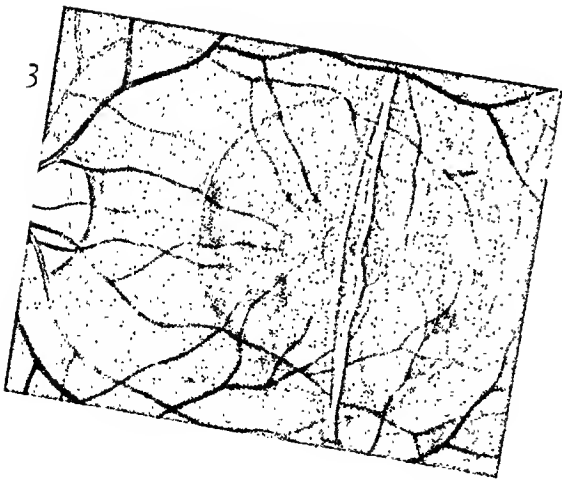
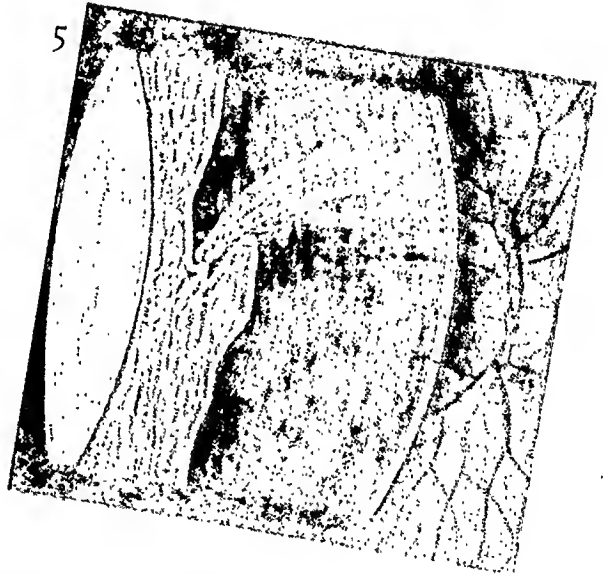
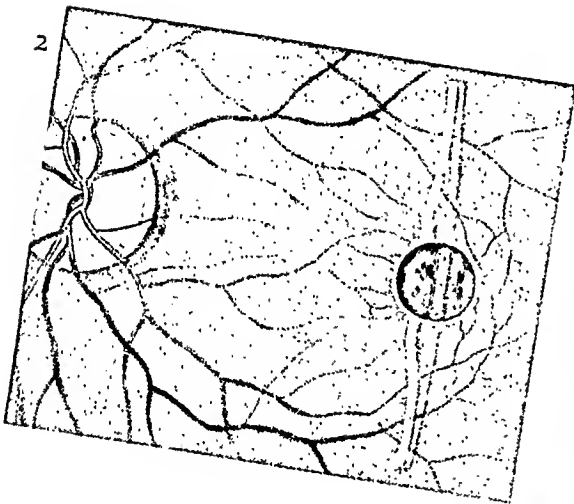
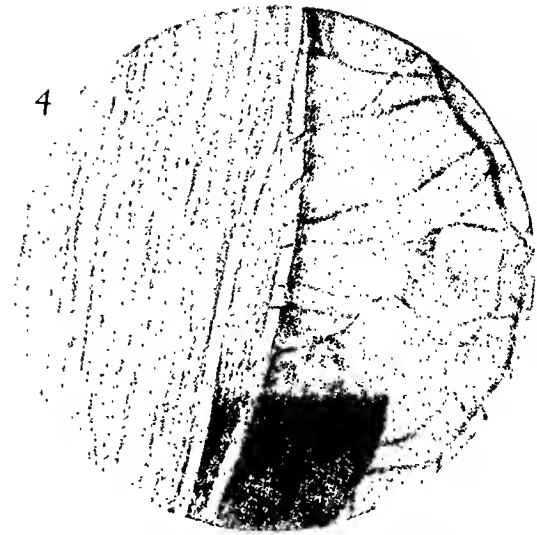
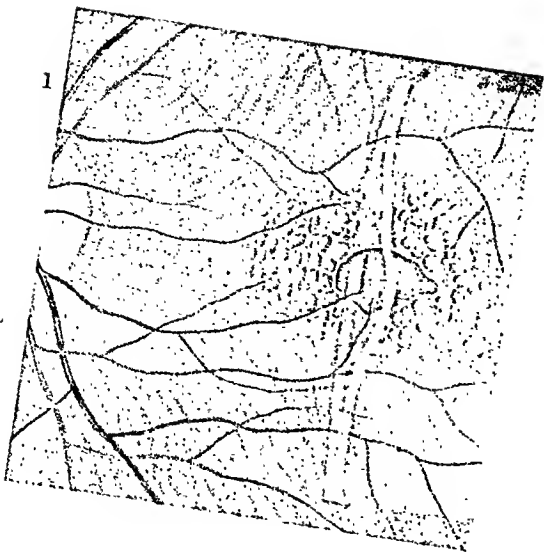
FIG. 3. Macula changes. Choroiditis serosa centralis. Male 34 years of age, case referred to clinic with diagnosis of acute retrobulbar neuritis. Within two weeks vision reduced to 0.6. variable central scotoma especially for blue.

Slit-lamp: The narrow beam (light directed from the right) discloses that the retinal surface is slightly raised, but in the center, the foveola is seen just below the surface. The retina itself is transparent. Its posterior margin is unsharp. Beyond this is a clear space (optically empty). Many small yellowish points lie in front of this space. Later central edema disappeared.

FIG. 4. Changes following contusion. Pre-retinal hemorrhage; male 27 years old. Fell from a horse. Head and right eye were injured by helmet. Hemorrhage below macula shaped like bicorne "Napoleon's hat." Purtschers retinopathy. Vision 1. Small central scotoma. Vitreous hazy.

Slit-lamp: Remnants of blood in vitreous. In the macula region there is a detachment of the posterior limiting layer of the vitreous. In a sac formed by this detachment there is a bicorne shaped hemorrhage (pre-retinal). Above, there is a hazy fluid filled with cells and limited posteriorly by the sharp retinal line (Light

* Courtesy of Dr. Hans Goldmann.



directed from the left). No retinal edema. Above the line of hemorrhage there is a hazy fluid-like collection of fibrin, and phantom red and white cells. Two months later the vision was 1.

FIG. 5. Extreme vitreous detachment. Male 28 years of age who one year previously had periphlebitis.

Slit-lamp: The light is directed from the right. Notice that the retinal posterior limiting layer of the vitreous is displaced forward near the lens. A band of vitreous seen faintly extends towards the retina. Retinal line of illumination is intact.

FIG. 6. Recurrent vitreous hemorrhage. Male 55 years of age. Retinal detachment.

Slit-lamp: Flat retinal detachment. Below in the region of the detached retina there is an extended red line marking the hemorrhage. Above this line is a triangular retinal hole. The slit-lamp shows that the convex blood line demarcates the place where the vitreous is separated from the retina. Above the line where the retina and vitreous are still together there is a collection of blood in the form of horizontal stripes. The vitreous detachment has only advanced to the immediate neighborhood of the retinal tear. From this point on the retina and vitreous start being lifted together.

The mechanism of the causation of recurrent hemorrhages and tear formation is as follows: The vitreous detachment starts as usual above. In doing this it damages the retinal vessels causing hemorrhages especially in older persons. It is a common observation that retinal detachment may be initiated by vitreous hemorrhages. As the vitreous detachment progresses it may cause small hemorrhages and finally produces a tear in the retina below at a time when the vitreous detachment has already far progressed.

so-called "physiologic" remains on the posterior capsule, these are rarely pigmented (Fig. 485).

Pigmented Stars. Both Streiff⁶²² and Collivati have reported the



FIG. 485. Striate and linear figures on the posterior lens capsule with unusual attachment of the rests of the hyaloid system.

finding of pigmented stars on the posterior lens capsule like those more commonly seen on the anterior lens capsule. However, these stars were associated with other anomalies (coloboma iridis, aplasia of the posterior retinal layer of the iris, and congenital subluxation of the lens). In two of Collivati's cases, the pigment stars were found anteriorly and posteriorly at the equator. Pigmentation of the zonular fibers as well as the vitreous occurred in one case.

Retrolental Fibroplasia. Attention was called to a massive growth of embryonic connective tissue in the meshwork of a persistent tunica vasculosa lentis by Terry (1941).⁶³² It is associated with prematurity. Normally, during fetal development, regression of the tunica vasculosa lentis occurs at 8½ months. Hence in prematurely born infants these vessels are present. Terry⁶³³ suggests that as a consequence of light stimulation fibroplasia occurs in the exposed vascular meshwork (Fig. 486). This is seen as an opaque vascularized tissue behind the crystalline lens in which the vessels tend to radiate from a central point. His theory stresses the motor action of light in that "active pupillary responses could embarrass the venous drain-

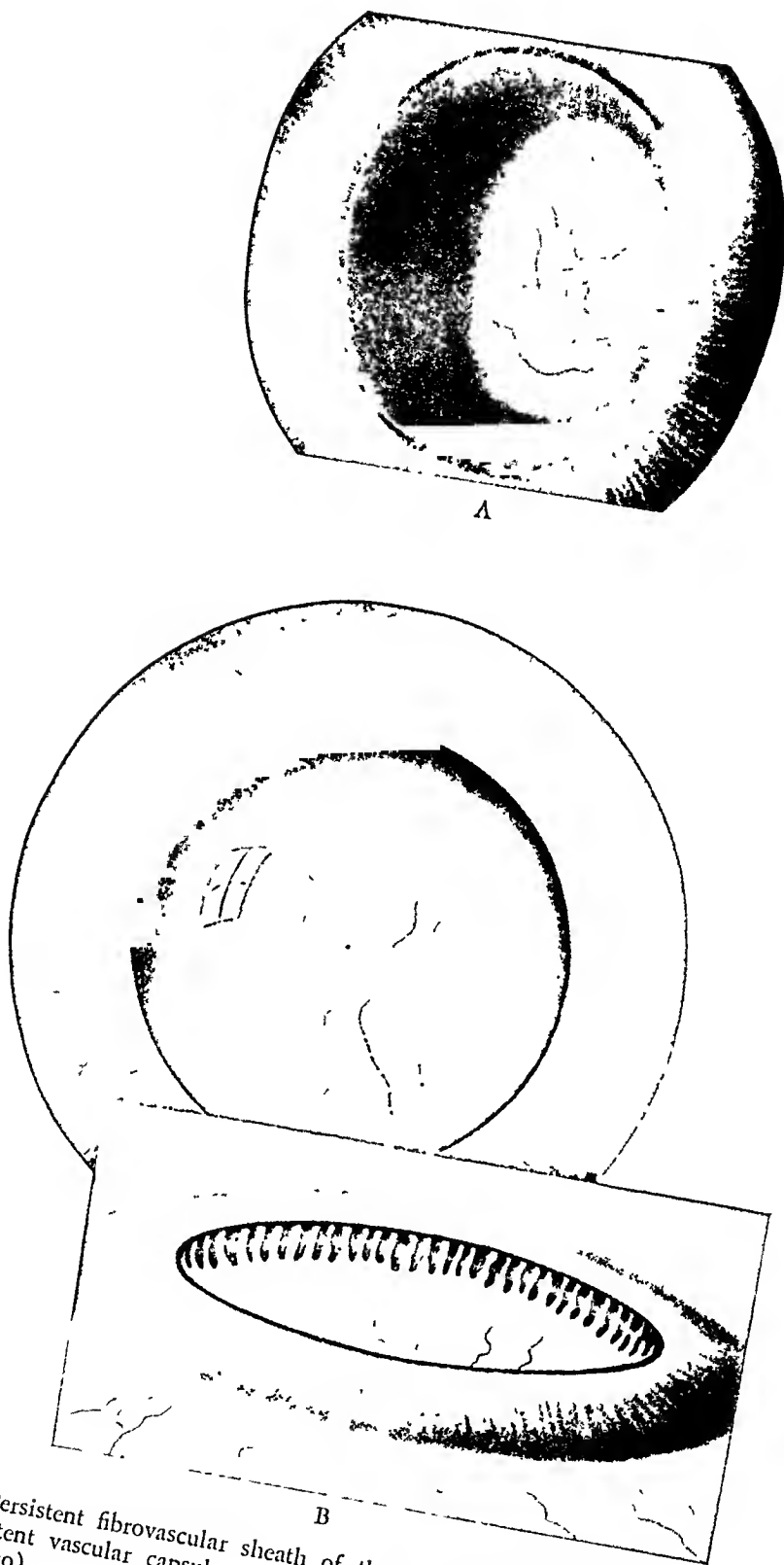


FIG. 486. A. Persistent fibrovascular sheath of the lens. Nine days old; right eye (after Reese). B. Persistent vascular capsule of the lens (pseudoglioma). Age, 5 months (after Reese and Troncoso).

age of the tunica vasculosa lentis by stretching and kinking the vessels as they extend from the posterior surface around the pupillary margin to the anterior surface of the lens." "Passive congestion in

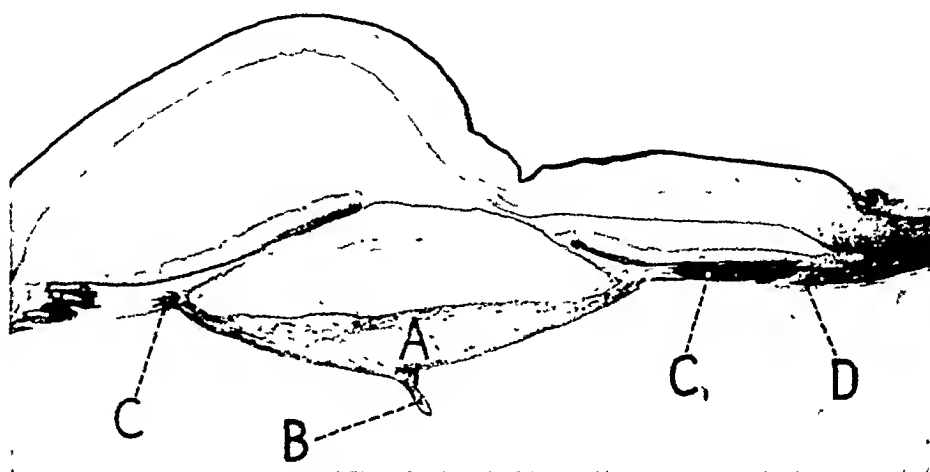


FIG. 487. A portion of the human eye showing persistence and hyperplasia of the primary vitreous corresponding to Clinical Type I. The retrolental fibrous mass is shown at *A*, the hyaloid artery at *B*. The lens is small. Ciliary processes, *C* and *C*₁, extend to the fibrous mass and the retina is seen as far up as *D*. (Reese.)

itself is not presumed to cause fibrosis, but the edema associated with it can produce fibrosis such as is seen in the so-called pulmonary osteo-arthropathies."

It has been noted that contraction and partial resolution of the fibroplastic tissue may occur when further development of the eye takes place postnatally provided no untoward complications arise (hemorrhage, glaucoma, and posterior synechiae).

In summarizing his discussion Terry⁶³⁴ stated: "Over 10 per cent of infants born very prematurely, weighing 3 pounds or less at birth, can be expected to be blind from retrolental fibroplasia. . . . The typical characteristics of the disease are opaque vascularized membrane behind the lens, microphthalmia, shallow anterior chambers, fetal-blue color of the iris, thin ciliary processes in front of the opaque tissue, searching nystagmus, apparent photophobia, persistent hyaloid artery, and often retinal separation."

Reese and Payne* reporting on 50 cases believe that retrolental

* Reese and Payne, *Am. J. Ophthalm.*, Vol. 29, No. 1, January 1946.

fibroplasia represents only one type of abnormality of the primary vitreous, and they prefer to designate the lesions as "persistence and hyperplasia of the primary vitreous." They state: "Terry employed the term 'retrolental fibroplasia' to designate primarily a condition occurring bilaterally in premature babies after birth (not earlier than four months). He felt that this is a new, acquired condition different from the lesion which is unilateral at the time of birth in full-term infants. From the study of our cases and the literature we do not believe such a distinction is justified but that the same lesion may occur in both premature and full-term babies, that it may be unilateral or bilateral, and that all are congenital but may not manifest themselves until sometime after birth either because they are not looked for or because the lesion progresses."

The reason for the apparent increase in incidence of this condition of late is probably related to the present lower mortality rate of premature infants. These authors divide the clinical manifestation into four types as follows:

- I. Saucer-shaped, whitish opaque tissue conforming to the posterior surface of the lens (Fig. 487).
- II. An opaque cornea, greatest in the central portion, usually associated with glaucoma, and maybe buphthalmos.
- III. A localized area of opaque tissue on the posterior surface of the lens, or at the lens equator, or in the anterior vitreous, with or without retinal detachment.
- IV. Remains of the hyaloid system.

"These four clinical types are not necessarily sharply demarcated but may merge. Type I may change into type II, or there may be type I in one eye and type II in the fellow eye. Otherwise one type cannot go over into another type."

From the clinical and pathologic findings it would seem "that the basic lesion is a persistence in part or in toto of the primary vitreous with or without hyperplasia and with or without secondary changes consequent to hemorrhage, opening of the lens capsule, and glaucoma."

Persistent adherence of the primary vitreous to the retina at the time when the secondary vitreous forms may lead to retinal separation.

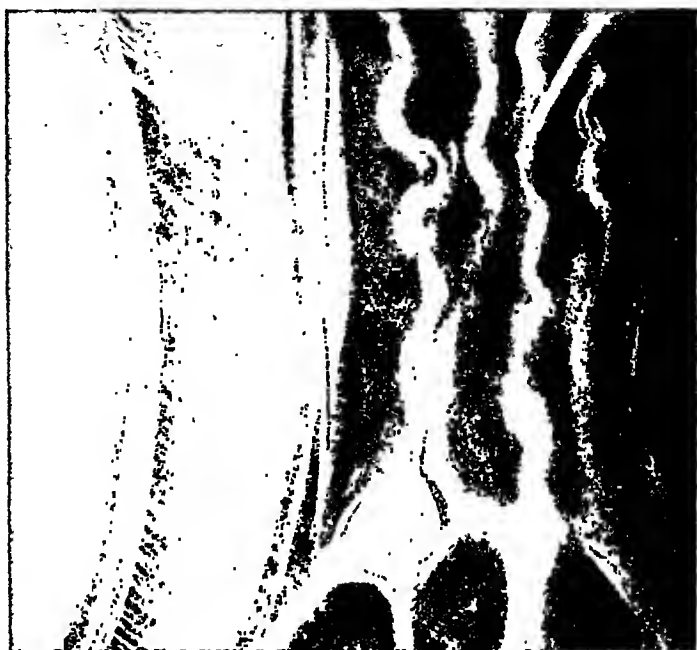
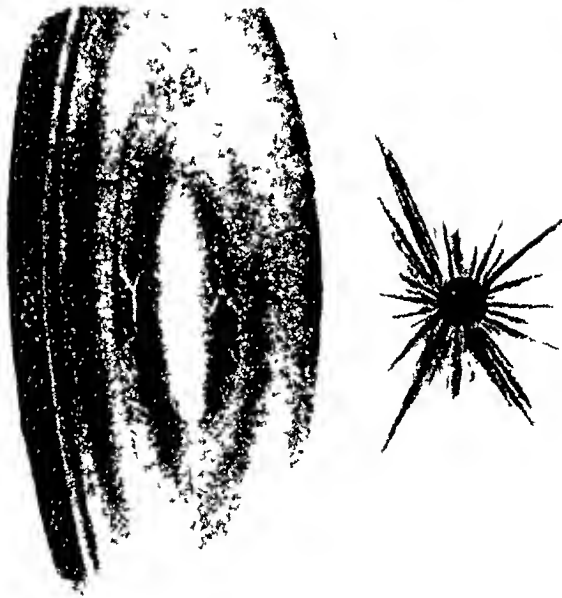


FIG. 488. Freely floating cystlike body in the anterior vitreous. Noninflammatory vitreous opacity. Female, 28 years of age, vision 0.7. Opacity was seen ophthalmoscopically. It had very little motility and was close to posterior lens capsule. Lens was normal. Hyaloid vitreous was of appreciable optical density, with fine oblique striations. There were several tracts of low luminosity, some of which converge toward the larger intense opacity. The opacity was situated below the arcuate line which is not shown. (After Koby.)

At the October, 1948, session of the American Academy of Ophthalmology and Otolaryngology William C. Owens and Ella M. U. Owens presented an interesting study on retrolental fibroplasia in premature infants observed in routine examinations in the Johns Hopkins Hospital. During this study they observed the onset, course, and development of the final picture of retrolental fibroplasia. None of the infants they observed had retrolental fibroplasia at birth. The disease was not related to persistence of the hyaloid system. Early postnatal examinations of the eyes revealed normal fundi. The earliest change observed was the formation of progressive, angiomatous dilations of the retinal vessels, followed by the development of extensive, grayish retinal edema and detachment. The vitreous became cloudy. The retrolental membrane was formed

A



B

FIG. 489. A. Cystic deformation of hyaloid artery. Eight-year-old child. Pearlike deformity of hyaloid artery. V. 0.6. Pearlike remnant inserted on the posterior lens capsule by a network of vessels. No arcuate line. Hyaloid vitreous is dark, with faintly visible striations. Anterior band of definitive vitreous is clearly seen with whitish lozenge-like forms separated by darker spaces. The same is repeated deeper but less clearly defined and less luminous. Two years later the pearlike remnant seemed smaller. (Modified after Koby.) B. Cyst; detached hyaloid corpuscle. A cystlike opacity of the vitreous—possibly a detached hyaloid corpuscle. Radiating fibers are probably remains of vessels. Small dark band extending upwards was reddish brown in color. This may be blood in the vessel.

by the extension across the retrolental space of fusing peripheral portions of the swollen, grayish retinal detachments. The vessels in the retrolental membrane had their origin from retinal vessels and not from vessels associated with the embryonic vascular sheath of the lens. In most cases the active stage of the disease subsided gradually. In some eyes the activity subsided before a complete membrane was formed behind the lens. Usually in these eyes bands simulating retinal folds extended through the vitreous to localized areas of retinal detachment. Newly formed vessels arising from the retina were visible in the folds. From their observations on the course of the disease they have been led to the conclusion that important and as yet unknown etiologic factors must be at work during the postnatal life of the infant. They felt that further work should be directed toward a study of the factors operative during the postnatal life of these infants.

Congenital Vitreous Cysts. Tansley (1899) first reported a case of vitreous cyst. Since then others have been described, apparently of noninflammatory or nonparasitic origin. Some attributed their origin to the ciliary processes. Troncoso (1903)⁶⁴⁰ reported a case in a young girl in whom an oval pigmented semitransparent cyst was seen in the anterior layers of the vitreous. Pigmentation of the surface of a cyst may be a secondary phenomenon and does not justify in itself the assumption that the cyst is of uveal derivation. Recently several cases of vitreous cysts (attached to the posterior lens capsule or freely floating in the anterior vitreous) have been examined biomicroscopically. According to the observations of Lampert and Koby, it appears that freely floating cysts can be derived from the hyaloid artery, even if no evidence of abnormal persistence of the vessel is found (Fig. 488). In one of Koby's cases, a whitish pear-shaped cyst was located at the insertion of the hyaloid artery. The vitreous was normal (Fig. 489 A). In another a pigmented cyst was seen attached to the posterior lens capsule above the insertion of the hyaloid artery, 2 mm. nasal to the posterior pole. The hyaloid vestige hung down behind the capsule undisturbed. He considered that the

cyst had detached itself from the vestige. Figure 489 B illustrates another form which I observed deeper in the vitreous.

Practically all the cysts described were monocular. Litinsky

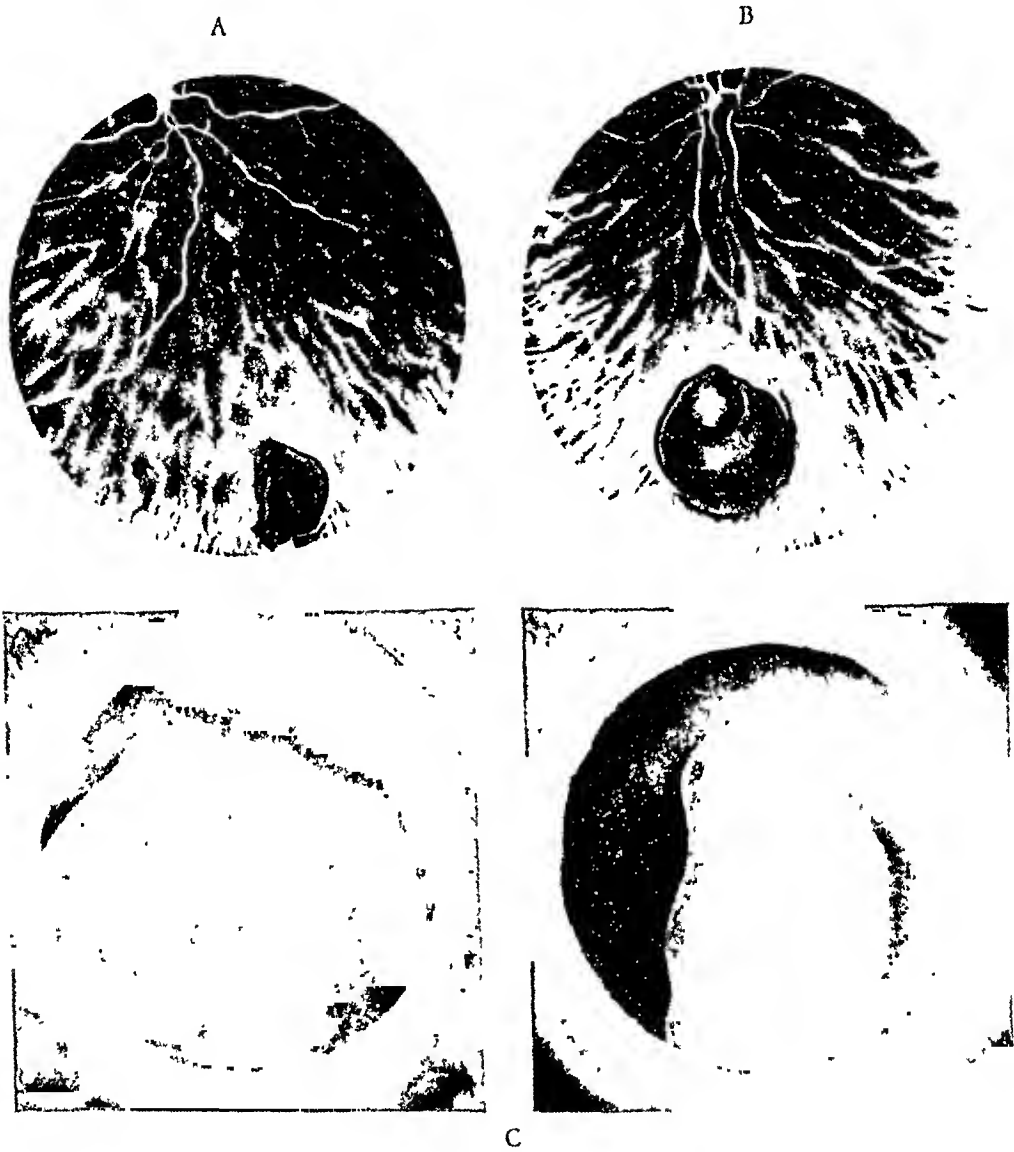


FIG. 490. Cyst of the vitreous. A. Right eye. Cyst of the vitreous cavity. J. B., age 54. Retinitis pigmentosa. B. Left eye. Cyst of the vitreous cavity, in the same patient. Retinitis pigmentosa. C. Ophthalmoscopic view of the above cysts (Perera).

(1931)⁵²⁶ and Perera (1936)⁵⁰⁰ have described bilateral cysts in retinitis pigmentosa.

In Litinsky's case there were bilateral slightly mobile spherical formations which, with the biomicroscope, had a reddish reflex. They were suspended by fine filaments which lost themselves toward

the region of the ora serrata. In Perera's case, there was a cyst in each eye associated with pigmentary degeneration of the retina (Fig. 490).

Perera states: "Behind the left lens, floating freely in fluid vitreous, was a large yellowish gray globular translucent structure, measuring from 3 to 4 mm. in diameter. Its upper pole was elongated and somewhat indented. The surface of this cyst was smooth, glistening and avascular and contained no pigment. No strands connecting the body with other intra-ocular structures could be identified. On elevation of the eye, the cyst floated up or 'bounced' into the optical and visual axes, obstructing the fundus reflex viewed through the pupil. When the eye was motionless, the cyst sank to the dependent portion of the vitreous cavity.

"The interior of the right eye revealed a similar but smaller cyst, which was somewhat oval.

"The patient was aware of a momentary blurring of vision when elevating his eye.

"During six months of observation, there was no change in the appearance or size of the cysts."

Chapter Thirty-Two

BIOMICROSCOPY OF VITREOUS OPACITIES

GENERAL CONSIDERATIONS

IN REVIEWING the older or prebiomicroscopic conceptions of vitreous opacities Roemer (1906) divided them into four groups: (1) physiologic muscae volitantes, (2) dustlike elements in the vitreous, (3) filaments and flocculations, and (4) membranes. The latter three may be seen ophthalmoscopically.

Muscae volitantes are those which cannot be perceived with the ophthalmoscope but only by the patient himself. Possibly they are incorrectly termed "physiologic," since in the light of present knowledge it is felt that they may be associated with pathologic (degenerative or abiotrophic) intra-ocular alterations. The dustlike elements are just visible with the ophthalmoscope especially if a plane mirror and a convex lens is employed (mirror loupe). In this way they can be made to project shadows and with movements of the eyeball to show the characteristic floating or oscillatory motions. The larger filaments, flocculations, and membranes are seen more easily. The farther forward the opacities are, the less they are able to project shadows, and hence they disturb the patient less. Those located deep cast shadows posteriorly upon the retina and consequently are more annoying. This probably accounts for the subjective improvement frequently noted by the patient, i.e., when deeper opacities make their way more anteriorly.

Ophthalmoscopically, it is permissible to assume that the vitreous is altered only when opacities are seen. Against the red glow of the fundus, the vitreous appears optically empty. As the illumination with the ophthalmoscope affords a means of illumination comparable only to transillumination or diaphanoscopy, opacities of suf-

ficient density will appear black or if crystalline, yellowish (Plate LXXX, figs. 6, 7, 8). Accordingly, with this method it is possible to see alterations only when they become well advanced.

With the focal beam of the biomicroscope the tyndall effect discloses any altered homogeneity or disarrangement of the fibrillae or lamellae forming the vitreous structure. These changes can occur with aging or in myopia. Although these alterations cannot be visualized ophthalmoscopically, from the standpoint of biomicroscopy they must be considered as opacities. The exact degree of density required to cause a biomicroscopically visible alteration to become apparent as an opacity with the ophthalmoscope is not known.

With progressive derangement or degeneration of its framework relatively dark spaces or lacunae form (liquefaction ?).^{*} (See Fig. 491.) Breakdown, agglutination, or condensation of fibrillae (microfibrillary degeneration) probably in the beginning results in the formation of muscae volitantes and eventually, when the formations are large and sufficiently dense, form the so-called floating opacities which can be detected with the ophthalmoscope (Fig. 492). Regarding the membranous types of opacities, it must be remembered that those more or less fixed in the axial regions may be congenital rests, i.e., related to the hyaloid system.

In addition to derangement of the framework and the formation

^{*} The existence of liquefaction (synchysis) of the vitreous was assumed originally because of the apparent free movement of visible opacities within it. Ophthalmoscopically it produces no detectable signs. Since freely floating opacities were seen in many conditions (myopia, senility, trauma, and inflammatory states) it was held that liquefaction had occurred. As Koby pointed out certain writers on the one hand state that liquefaction is preceded by the formation of opacities and on the other that even a very liquefied vitreous can remain very transparent; that in the end one can only recognize liquefaction by the presence of highly mobile opacities. At best this leaves us on very uncertain ground. The physicochemical basis of this change is thought to be one of dehydration of the vitreous micellae, resulting in a condition in which absorbed intra-ocular fluid (water?) became separated from them. According to Duke-Elder,⁴¹¹ "It is due to a conversion of colloidal gel into a sol and is usually associated with development of ophthalmoscopically visible opacities formed by the colloidal micellae which aggregate together in dustlike particles, strands or membranes." In this case clear spaces (water) should be found between the remaining distorted and agglutinated trabecular structures. Actually with the biomicroscope (arc illumination) no optically clear (dark) spaces—indicating the presence of pockets of fluid are found. Dark spaces are seen with light sources of low intensity but with the arc lamp (similar to the normal retrolental space) these spaces become relucant owing to the presence of a trabecular structure. Evidently the physicochemical concept of liquefaction does not exactly coincide with clinical findings. It may be that a relative decrease of viscosity takes place rather than the formation of water-clefts, giving the impression of vitreous liquefaction.



FIG. 491. Distorted framework of the vitreous in myopia. Female, —14 D. Note large dark areas (optically empty). Greater part of the framework is composed of membranes and threads that oscillate freely with the eye movements. With cessation of eye movements these structures do not gravitate but tend to return to their original position of rest. Luminous threads seen anteriorly are probably remnants of the vasa hyaloidea propria. (After Vogt.)



FIG. 492. Senile opacities in the vitreous. Female, age 65. Ophthalmoscopically, floating opacities are seen. Hyaloid vitreous is not sharply differentiated from the definitive vitreous. Some microfibrillary degeneration is seen anteriorly. The whole vitreous is constituted of irregular fibers of variable thickness and lengths. In two places the fibers are condensed so as to form large cottonlike fluffs. Large empty spaces occur near condensations. These condensations are probably muscae volitantes. (After Kobay.)

of dark lacunae, biomicroscopically destruction is signalled by a dustlike or flaky cloudiness and frequently by the presence of white, cottony fluffs (Fig. 492). It is interesting to compare the ophthalmoscopic appearance of a vitreous undergoing trabecular degeneration, as indicated by large floating opacities (e.g., in myopia) with what is seen biomicroscopically. The beginner will often be chagrined at his inability to identify a large floater so easily seen with the ophthalmoscope. This results from the difference in illumination employed. In the first place if the opacity is too far back (beyond the anterior one-third) the beam may not be able to reach it. Secondly, even if the opacity is anterior, passage of the beam into the vitreous will reveal a very nonhomogeneous vitreous with large dark spaces and irregular sinuous and distorted grayish bands (condensed trabecular tissue) and fibrillary degeneration. As one observes this type of tyndall effect, it is usually not possible to identify the exact place or structure which ophthalmoscopically is dense enough to produce a shadow. This brings to mind the difference between the appearances of certain structures in the cornea and lens when viewed by direct focal light and by retro-illumination (a form of diaphanoscopy) (Vol. I). Although we are able to see features within the tyndall phenomenon that are invisible in diffuse or reflected light, unless the differences in index of refraction between the feature and its surroundings are very great, actual contrast within the tyndall beam itself may not be as marked as the contrast obtained by retro-illumination (shadow effect). In lieu of special apparatus designed for this purpose (page 1380) opacities in the deeper parts of the vitreous (even those located near the fundus) may be seen with the plane mirror and loupe (indirect ophthalmoscopy) especially if the light source is intense. These may escape observation with the ordinary electric ophthalmoscope.

Increases in relucency result when a difference occurs in the index of refraction between the fluid of the vitreous and the framework. Normally these indices are practically the same. It should be emphasized that these changes ordinarily are not seen with the ophthalmoscope unless they are pronounced, and that they result from

changes endogenous to the vitreous structure, in other words, not from cellular infiltration, e.g., blood cells, fibrin, or uveal pigment. Cellular infiltration (exogenous) in the vitreous forms another class of opacities. When fairly dense, they cause the ophthalmoscopic view of the fundus to be hazy. The deposition of cells may occur without biomicroscopic alteration in the vitreous framework and are capable of absorption. Depending on the gravity of the condition both trabecular destruction and cellular deposition may coexist.

According to biomicroscopic appearances, Koby has listed vitreous changes (opacities) in the order of their seriousness: *

1. A diffuse increase in intensity or relucency of the tyndall effect. This picture is also often produced by trauma and may be ephemeral.
2. Disseminated cellular deposition (tiny dotlike opacities) without any apparent alterations in the framework. They are seen in the uveitides, sympathetic ophthalmia, etc.
3. Alterations of the framework — fibrillary and membranous (lamella) degeneration. This is seen especially in myopia and in senility.
4. Combined forms, where destruction of the framework is associated with cellular deposits of hemorrhagic or inflammatory origin.

The first two can only be observed with the biomicroscope; indications of the second and third may be found ophthalmoscopically

* Etiologically, Duke-Elder ⁴¹² divides vitreous opacities in three classes:

- I. Congenital remnants of the hyaloid vascular system
- II. Endogenous opacities
 - A. Protein coagula of the colloid bases of the gel;
 - B. Crystalline deposits
 1. Asteroid bodies
 2. Synchysis scintillans
- III. Exogenous opacities
 - A. Protein coagula — the plasmoid vitreous
 - B. Exudative cells
 - C. Blood
 - D. Tissue cells: epithelial, histocytic, glial
 - E. Tumor cells
 - F. Pigment; melanotic and hematogenous
 - G. Parasites

It should be noted that biomicroscopically, it is not possible to identify or differentiate certain of the opacities listed under exogenous opacities, e.g., tissue cells. In my experience a purely plasmoid vitreous does not occur without exudative cells.

when they are very marked. The exact appearance of all these alterations, even in conditions of the same etiology, varies from case to case. This is not surprising when one considers the pleomorphic aspects even of the normal vitreous. Individual variations and appearances of opacities will be considered in the chapter on pathologic changes. However, as already indicated in the consideration of cellular deposits in iritis (page 823), caution must be used when judging the color of cells deposited on the vitreous framework.

SENILE AND MYOPIC ALTERATIONS

The alterations observed in the vitreous biomicroscopically are so similar in senescence and in high degrees of myopia that they will be considered together. This similarity was first stressed by Vogt (1924) who pointed out that the vitreous changes in the two conditions, viewed biomicroscopically, often defy differentiation.* Fundamentally the alterations are of a degenerative nature and are characterized by a disruption of the vitreous framework with resulting production of endogenous opacities (without cellular infiltration). Cellular infiltration results from inflammatory and hemorrhagic processes, and hence its presence or absence is valuable in differential diagnosis.

The concept of liquefaction (based on the presence of floating opacities and "dark" spaces) so often stressed in these forms of degeneration is, as already indicated, of uncertain validity (page 1402). Even in such cases as *synchysis scintillans* in which fluidity is regarded as a principal change, the biomicroscope may reveal little if any change in consistency. Hence, although one can conceive of a relative loss in consistency the expression "liquefaction" should be employed with reserve.

In old age and in myopia (depending on the degree and progressiveness) the normal, somewhat regular, arrangement of fibrillae and lamellae is lost (Fig. 491). Condensations in the disrupted vitre-

*Vogt struck a parallel between the vitreous alterations in myopia and senility to the degenerative changes occurring in the fundus in these same conditions (circumpapillary-foveal and peripheral degeneration). In both, the changes may occur abiotrophically in the young (presenile) as well as in older individuals.



FIG. 493. Senile vitreous alterations. "Skein of wool" type of changes. Note twisting of fibrillae and condensations caused by agglutinations of fibers or thickenings at the places where they cross one another.



FIG. 494. Vitreous in myopia. Female, age 34, —20 D. Ophthalmoscopically the vitreous appears transparent. With the biomicroscope the retrolental space is dark and there is no clear demarcation between the hyaloid and the definitive vitreous. Anterior parts of the vitreous are fibrillar with nodular condensations. Deeper there are structures suggestive of pseudo-membranes. The nodular-like white condensations may be the rudiments of microscopic opacities. (After Koby.)

ous framework account for the vitreous opacities seen ophthalmoscopically (Fig. 492). In the presence of numerous condensations usually seen in the tyndall beam in these conditions, there is not always sufficient difference in density to identify any particular one with the opacity seen ophthalmoscopically. No correlation is found between changes in the vitreous and those in the lens in senility. Marked senile and myopic alterations in the vitreous may occur in the presence of a clear lens. After cataract extraction the changes in the vitreous can be quite minimal.

In the early stages, the vitreous may not exhibit any marked increase in relucency in spite of the disturbance in its architecture. However, with increases in condensation one does get the impression of loss of transparency as evidenced by a more pronounced tyndall effect. Such condensations occur in places where parts of lamellae or fibrillae become twisted or knotted together to form a "skein of wool" or dustlike opacification (Fig. 493).

The so-called "retrolental space" (primary vitreous) may gradually become occupied by distorted scaffolding so that finally larger irregular accumulations approach the posterior surface of the lens. It should be noted that there may be differences in behavior between the hyaloid and definitive vitreous and also within the definitive vitreous itself. This gives the impression of ill-defined successive zones within the tyndall beam. For example, in high degrees of myopia even in comparatively young individuals (20 to 40 years of age) one may first see with ordinary illumination that the retrolental space is still relatively dark (Fig. 494). Bordering this there may be a zone occupied by filaments of irregular caliber. Many seem to run in an irregular way but in the main they tend to hang vertically. Focusing a little deeper one gradually comes to an area occupied by diaphanous veil-like membranes of varying density. These veils seem to be suspended in a darker substrate. But with stronger light sources (arc) it will be observed that this dark substrate in turn becomes more reluctant owing to the presence of formations not hitherto seen. In places, an irregular fibrillation may be found on these veils or pseudo-membranes (microfibrillary degeneration). In the depths as far as one can see, the appearance is the same but the veils tend to

become more irregular and wrinkled. With movements of the eyeball all these formations are suddenly set in motion, causing momentary changes in their relations and aspects. In some cases, the



FIG. 495. Microfibrillary degeneration seen in a patient aged 80.

formation of veils may not be so marked in which case one gets the impression of a less distinct tyndall effect. In these cases, large, relatively dark spaces (especially in myopia) are enclosed by irregularly-coursing fibrillae. Occasionally as part of them there may be cottony or flakelike opacities — evidently condensation products. Mixed types may occur in which a veil may be seen coursing sinuously between fibrillae. Or the entire vitreous from the posterior surface of the lens may be occupied by veils of marked relucency so that no stratification can be observed. Evidences of microfibrillary degeneration may be seen in the deeper parts, in the form of short irregular lines* (Fig. 495). This type of fibrillary degeneration may also

* In a case of a patient aged 61 years, having senile lens changes, Koby described three zones in the vitreous tyndall effect. In the first, just behind the lens capsule it was just possible to make out oblique striations, especially marked below. The second zone contained filaments going in all directions showing white nodes. The third zone contained fine short, broken filaments, in the form of commas and corkscrews of varying luminosity (microfibrillary degeneration) (Fig. 496).

occur in the hyaloid vitreous in myopia in which the entire retrolental space is filled with short curved fragments resembling commas or corkscrews (Fig. 497 A). In order to see such delicate changes a



FIG. 496. Senile vitreous. Female, aged 61. Senile changes in the posterior cortex of the lens. Ophthalmoscopically the vitreous was negative. With the biomicroscope three different zones can be seen in the vitreous: 1, area of faintly marked oblique streaks below; 2, dark space containing long filaments, many having fine thickenings; 3, zone of microfibrillary degeneration, with curled fibrils. This last zone appears dusty. (After Koby.)

high degree of illumination and magnification as well as a certain degree of dark adaptation are required.

Koby⁵⁰² has described an inconstant change, senile "peppering" of the vitreous which when present is characteristic. "It takes the form of minute white irregularities on the broken strands, giving them an appearance of having been sprinkled with fine sugar. They appear to be formed also on the vestigial vessels, which are less easily differentiated from the fibrinous strands than in youth." Ordinarily the comparatively larger and whiter vascular filaments because of their regularity will not be confused easily with trabecular fibrillae (Fig. 497 B).

DETACHMENT OF THE VITREOUS (SENILE AND MYOPIC)

Since the middle of the last century, there has been considerable dispute as to whether or not posterior detachments of the vitreous

A



B



FIG. 497. A. Myopic alterations in the vitreous. Female, aged 51, —17 D. Ophthalmoscopically the vitreous shows a high degree of heterogeneity with pseudomembranous-like opacity. The hyaloid vitreous shows microfibrillary degeneration with many fine filaments of low luminosity, sprinkled in places with fine dots. Deeper there are pseudomembranes running obliquely, anastomosing and enclosing dark spaces. A fine striation is seen on the pseudomembranes. B. Senile peppering of the vitreous.

seen in histologic preparations are due to artefacts (postmortem and fixational). Lately, according to the findings of Samuels (1930)⁵⁹⁴ and von Sallmann (1936)⁵⁹⁰—the latter using “nonshrinking” fixatives—it would seem that many such detachments are actual. Clinically the ophthalmoscopic finding in myopia and senility of a delicate membrane hovering in the deeper parts of the vitreous and distinguished by a prepapillary hole (corresponding to the optic disk—area of Martegiani) caused several investigators (Briere, 1875; ³⁷⁰ Galezowski, 1877; and especially Weiss, 1885⁶⁷³) to suspect that this represented a separation of the posterior limiting layer of the vitreous.* The edges of the opening are usually somewhat cloudy so that looking through it the fundus appears clearer, as if it were seen through a window. In several of the earlier reports, the investigators were uncertain whether they were dealing with fenestrated vitreous membranes, congenital anomalies (rests of Cloquet’s canal), or with an actual detachment of the posterior limiting layer of the vitreous. Later Kraupa (1914)⁵¹⁰ revived interest in this subject and affirmed Weiss’s contention that, clinically, this picture signified a posterior detachment of the vitreous. It should be emphasized that not all vitreous opacities are derived from holes or tears of the posterior limiting layer. As pointed out previously, most of them are the result of disruption of the intravitreal framework.

More recently, Pillat,⁵⁶³ Comberg, Vogt, Lindner, Sallmann and Regen, and Kleefeld† showed that sudden detachment of the vitreous, formerly overlooked, is a very common occurrence in myopia and in senility. It may also be a sequelae of pathologic states. Apparently the strength of attachment of the posterior limiting layer of the vitreous in the area about the papilla is even less than that of the vitreous base (ora serrata). Vogt considered detachment of the vitreous and the resultant ring formation as characteristic of degeneration of the posterior parts of the eye in myopia and in

* Elschnig, Salzmann and Wagner have contended that the prepapillary ring substratum consists of glial tissue in part as evidenced by histologic studies. This view is not inconceivable considering the embryonic relationships of Bergmeister’s papilla.

† Kleefeld employed modern techniques: binocular ophthalmoscope, biomicroscope with mirror and contact lens, modified Zamenhoff apparatus.

senility. Thus, it could belong to the group of changes commonly seen in these conditions, e.g., circumpapillary atrophy, equatorial and macular degeneration of retina and choroid, etc.

The detachment usually occurs suddenly, the patient experiencing photopsia (flashes, sparks, etc.). This may last only for a few days and, if minimal, may not be reported or be considered alarming by him. It generally is followed by blurring of vision. This, if slight, may also pass unnoticed by the patient. Others frightened by these symptoms will seek advice. Ophthalmologists not conversant with this condition, after finding no evidence of recent changes in the fundus, have erroneously diagnosed it as vitreous hemorrhage in the old or as a circulatory disturbance in myopes. With the simple ophthalmoscope or better with the plane mirror and a lens of plus 6 to plus 9 diopters, the mobile prepapillary ring may be observed clearly.* In its most obvious form, the ring is located in front of the disk, its periphery being somewhat opalescent (Plate LXXX, figs. 1-5). The remainder of the detached membrane may not be clearly apparent, its presence being indicated by a veiling of the fundal reflex. Radial projections are often seen extending from the margins of the ring (expression of the radial structure of the posterior limiting membrane?). Movements of the ophthalmoscope will alternately cause the ring to become luminous or dark (shadow effect). With time the appearance and location of the ring changes. As it comes forward, it becomes more mobile and tends to lose its circular character. Also the patient, entoptically, becomes less aware of it. If the ring assumes a sagittal or horizontal position, the lumen may not be apparent and only its edge (appearing as a sinuous line) can be projected. In this case it may become difficult to differentiate the tear from ordinary endogenous intravitreal opacities. In addition to the circumpapillary tear and detachment, holes in other areas of the posterior limiting membranes may occur. These may not be prepapillary and frequently are multiple. This possibly occurs as a

* Both Kraupa and Vogt recommend the employment of strong sources of illumination with the indirect method of ophthalmoscopy—the arc lamp or red-free illumination (direct current) gives the best results.

PLATE LXXX

FIG. 1. Detachment of the posterior limiting surface of the vitreous. Ophthalmoscopic view.

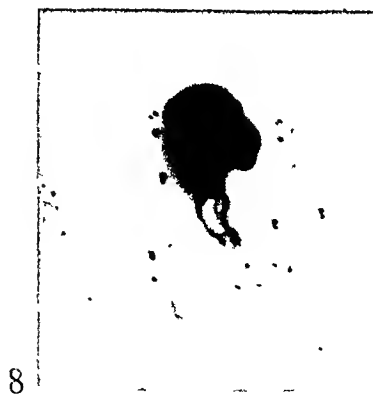
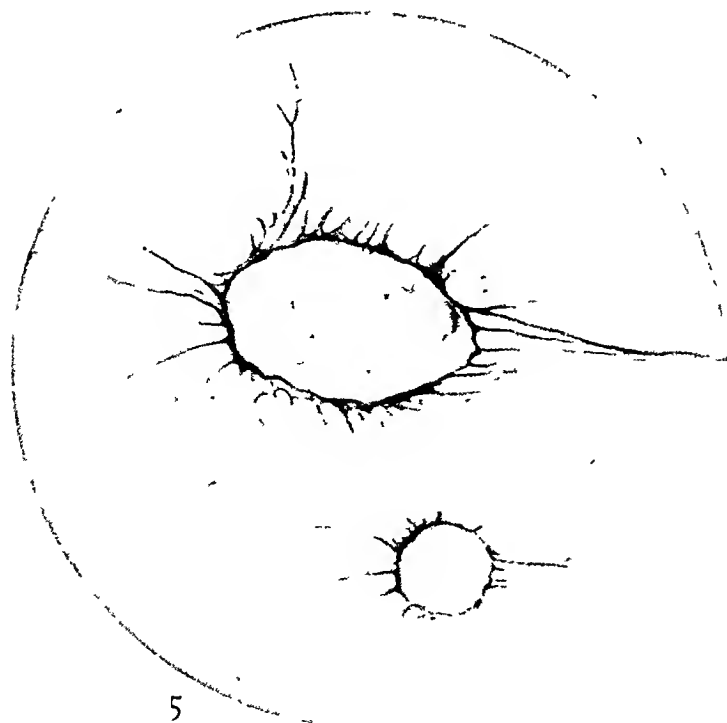
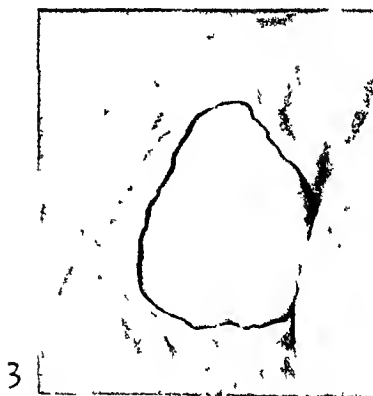
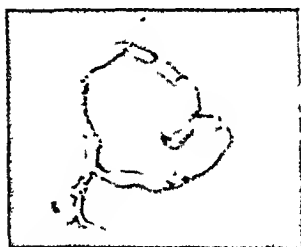
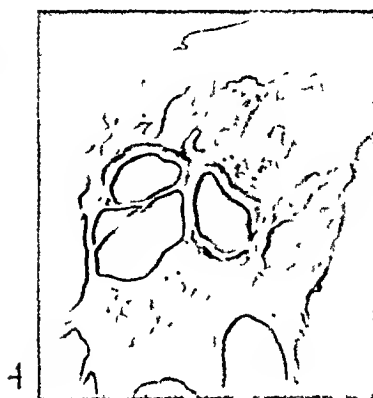
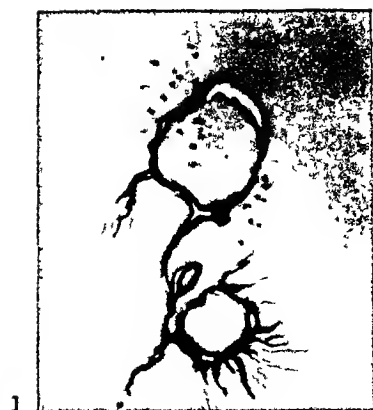
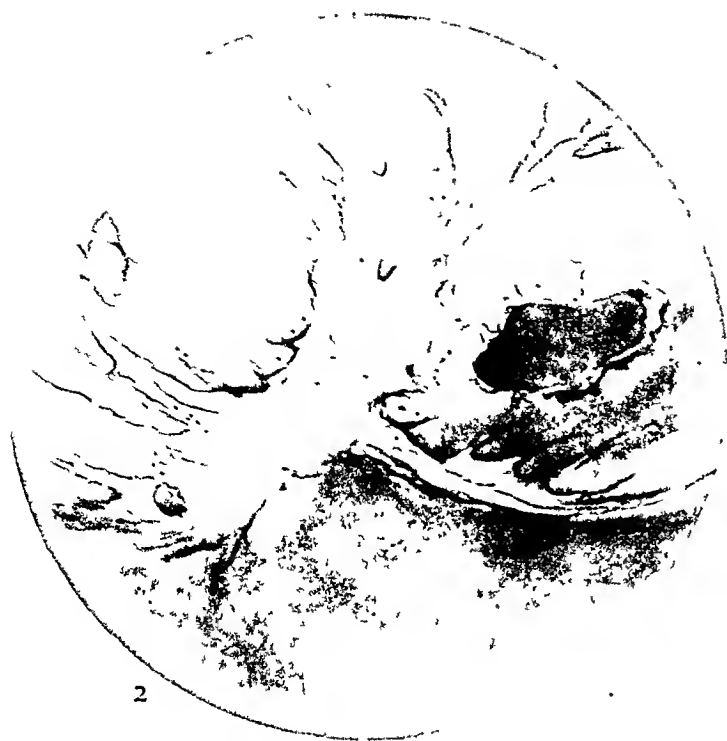
FIG. 2. Same case as in Figure 1. Showing several holes and surrounding opacification.

FIG. 3. Detail of the hole in the posterior limiting layer of the vitreous.

FIG. 4. Fenestrated type of detached limiting layer of the vitreous.

FIG. 5. Detachment of the posterior limiting surface of the vitreous. Showing details at the edges of the holes.

FIGS. 6, 7 and 8. Various forms of vitreous opacities as seen ophthalmoscopically.





result of more extensive retinal and choroidal changes. Vogt noted it in a case of choroiditis juxtapapillaris (Jensen).

For examination of the posterior vitreous, Lindner used a modi-



FIG. 498. Detachment of the posterior limiting layer of the vitreous. Observation by means of contact lens showing, left to right, retina, posterior limiting layer of the vitreous, lens and the cornea. (After Vogt.)

fied Koeppel contact lens. Kleefeld modified the Zamenhof ophthalmoscope by inserting a metal ring in its system (page 1383). Lindner,* Sallmann, Goldmann and Rieger, by means of the contact glass, demonstrated with the optic section preretinal detachments of the vitreous. Only by means of tyndall effect (narrow beam) is it possible accurately to visualize a membrane as delicate as the posterior limiting layer of the vitreous. However, in attempting to illuminate the deeper vitreous by any method, the red reflex from the fundus (diffuse illumination) always causes some interference (Fig. 498).

In many cases in which detachment of the vitreous is suspected, it is possible even without a contact lens later to demonstrate a superiorly detached posterior limiting membrane. It is preferable to observe the passage of the beam with the unaided eye (macroscopi-

* Lately Lindner has recommended the Hruby glass (see page 1386).

cally — the method of fentoscopy). In this way, with the 10-cm. illuminating lens and without any other special aids one can follow the vitreous tyndall effect to a depth corresponding to three or four

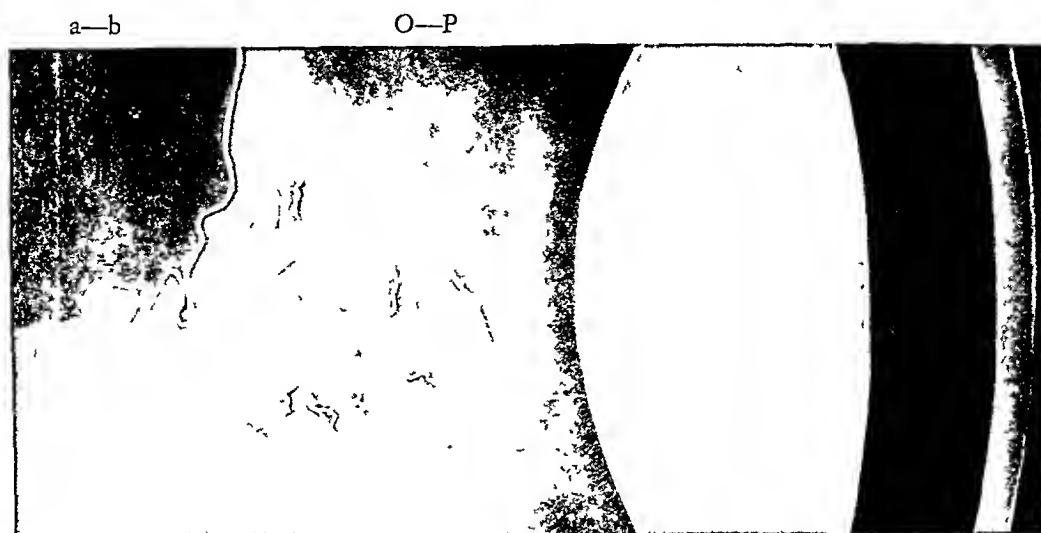


FIG. 499. Detachment of the posterior limiting layer of the vitreous. (After Vogt.)

lens thicknesses. It frequently happens that the detached and anteriorly retracted posterior limiting layer falls within the zone illuminated by the beam (Fig. 499). This membrane will be seen as a mobile structure limiting the contained vitreous structure which, owing to gravity (during moments of rest), assumes a dependent position. Above this, a clear (dark) area indicates that it is free of vitreous framework. With slight movements of the eye the limiting membrane and its contained structures beneath it are set in motion causing a change in the relationship between them and the darker (optically free) area above them. With rest the original picture is resumed. At first, for example, the zone *a — b* is small and limited to the dorsal part of the vitreous but with progression the retrolental portion *O — P* may be reduced to a thickness comparable to that of the lens (Fig. 500). The nature of the contents of the relatively dark area behind or above the detached posterior limiting layer and its contained trabecular structure is not known exactly. One should like to compare it to the postretinal fluid in retinal detachment which is slightly viscid (not like aqueous). In any event, there is sufficient difference in index of refraction between the detached frame-

work-containing part and the dark part to cause a significant lack of tyndall dispersion in the latter.

Comberg, Koby and others have described the rarer anterior de-



FIG. 500. Detachment of the posterior limiting layer of the vitreous. (After Vogt.)

tachment of the vitreous especially in trauma or after inflammatory conditions. Many of the investigators considered that detachment of the vitreous was always associated with or at least was a precursor of "spontaneous" retinal separation, and hence was the cause of it. The fact that detachment of the posterior lamella of the vitreous occurs so commonly and that in a few reported instances only was it followed by retinal separation, would indicate that no direct causal relationship exists between them. However, an exception to this may be true when instead of a "clean" separation of the posterior limiting layer of the vitreous, tags or strands of vitreous remain attached to the retina. Traction of such strands to a preexisting degenerated retina (see *infra*) might conceivably start the formation of a retinal hole. Biomicroscopically, in spontaneous detachment of the retina fixed membranes (inflammatory) have not been found in the vitreous. Some authors have held that these strands might by contraction detach the retina. There is considerable evidence today that a retinal tear or hole formation is a *sine qua non* of spontaneous retinal detachment. Histologic examination (Hansen-Kümell, Sourdille, and Kronfeld) in cases of recent detachment

(after 7 to 60 days) revealed no indication of any inflammatory process. Peripheral chorioretinitic (cystic) degeneration and atrophy was seen in the neighborhood of the tear. Only fresh cases of retinal detachment, i.e., before secondary pre- and postretinal changes have occurred, can histologically give reliable information. Vogt (1934)⁶⁶⁴ confirmed this and in addition found in some of his cases, an "uncapped" cyst. In other words, a peripheral retinal cyst in which its surface (facing the vitreous) was partly unhinged. Vitreous framework was adherent to this partly detached cap. According to Vogt retinal (noninflammatory) cystic and atrophic degeneration forms the basis of hole formation and subsequent retinal separation. Clinically, the detached cap of retinal tissues or fragments thereof have been observed floating in the vitreous or adherent to some of its filaments. Evidences of this have been seen ophthalmoscopically, not necessarily coincident with actual retinal separation in every case. The actual hingelike uncapping (hole formation) is a consequence of the swirling about (*jactatio corporis vitreii*) of an altered vitreous, attachment of which normally is probably firmer in the region of its base at the ora serrata. Vogt's hypothesis that spontaneous retinal detachment is primarily dependent on retinal degeneration and only secondarily on vitreous degeneration could also be valid in the nonmyopic cases or senile cases. Myopic lengthening of the globe is not contributory by itself to retinal separation, as has hitherto been thought by many writers; actually, like senile degeneration, it merely contributes to eventual separation. The same principle may obtain in postoperative detachment of the retina following discission or extraction of secondary cataracts (membranes), which may be due to traction upon a susceptible retina transmitted by adherent vitreous strands. In older persons, detachment of the retina may be accompanied or preceded by a sudden and massive vitreous hemorrhage. This may prevent ophthalmoscopic visualization of the fundus at the time the patient is first seen. After a week or 10 days absorption of the blood permits the detection of a retinal separation. In 3 of my cases recently observed, the retinal separation occurred in the region of the upper temporal quadrant. All had tears. In one

case the tear was found directly in the path of a small retinal vein, giving the impression that it had been torn across. All three were cured by operation.

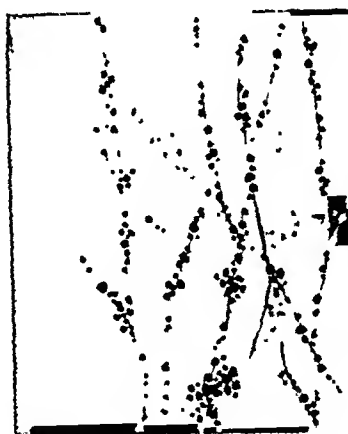


FIG. 501. Asteroid hyalosis.

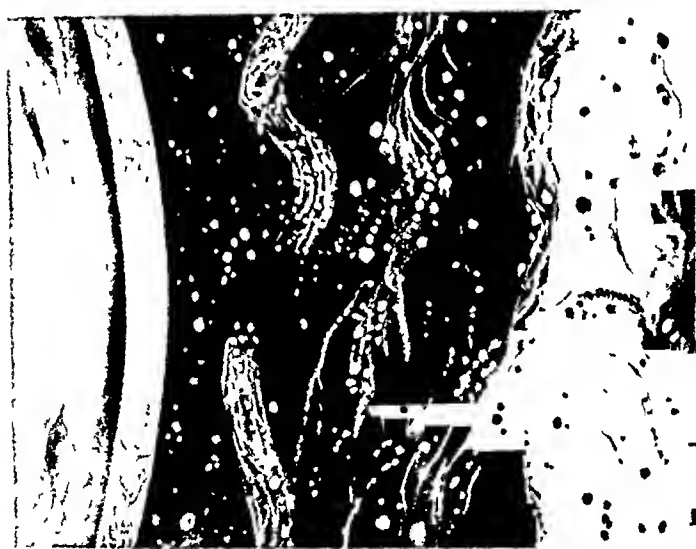


FIG. 502. Asteroid hyalosis. Note pseudomembranes. Male, age 71, vision 0.5. Ophthalmoscopically, besides brilliant opacities, pseudomembranes appeared like spider webs. With a biomicroscope the whole of the vitreous showed round brilliant opacities which extended nearly up to the posterior lens capsule. There were many filamentous and membranous formations resembling spider webs. These were encrusted with round opacities of varying sizes—all very luminous. The lens shows early signs of posterior cortical cataract. (After Koby.)

SYNCHYSIS SCINTILLANS AND ASTEROID HYALOSIS

Considering the crystalline nature of the deposits in synchysis scintillans and the calcium soap composition of those in asteroid "hyalitis," it would appear that, strictly speaking, their endogenous

nature is open to question. However, in view of the fact that these changes may appear in the absence of any other clinically demonstrable deteriorations in the eye most authors have preferred to classify them as strictly degenerative changes in the same sense that myopic and senile changes are so classified.

Asteroid Bodies ([Asteroid "hyalitis" or hyalosis]; scintillatio albescent [Benson's Disease]): This unusual type of deposition not uncommonly seen in the older age groups, usually in the seventh and eighth decades, presents a very striking biomicroscopic and ophthalmoscopic picture. In a large proportion monocular involvement is the rule. They do not have any significant pathologic portent. Ophthalmoscopically, in the glow of the reflected light from the fundus asteroid bodies appear yellowish-white, shiny "looking like snowballs or stars in the night sky."⁴¹⁰ Their shape is spheroidal or oval. They are commonly disseminated throughout the vitreous but also may be seen in localized strands or clumps arranged in columns. With the biomicroscope one is struck by a picture of unexcelled beauty. They appear as matt-white disks, spheroids and ovoids which in some instances gleam brilliantly (Fig. 501). There may be very little disturbance in the vitreous framework. However, at times a slight alteration may be noted in the form of poorly luminous filaments or networks arranged like spider webs and covered with a brilliant dust (Fig. 502). With eyeball movements these bodies oscillate only slightly and quickly return to their original position. This point differentiates them from the more mobile crystalline deposits of synchysis scintillans. As the beam traverses the vitreous, those directly in its path and in clear focus shine brilliantly whereas those to one side appear dimmer and larger because of the effect of diffusion circles (Fig. 503).

It is only recently that the differences between asteroid hyalitis and synchysis scintillans have been stressed (see Vol. I). Synchysis scintillans although rarer tends to occur in younger individuals and is more often bilateral. In contradistinction to asteroid "hyalitis" the particles in synchysis scintillans, which are crystalline and angular rather than rounded, float about with greater rapidity and

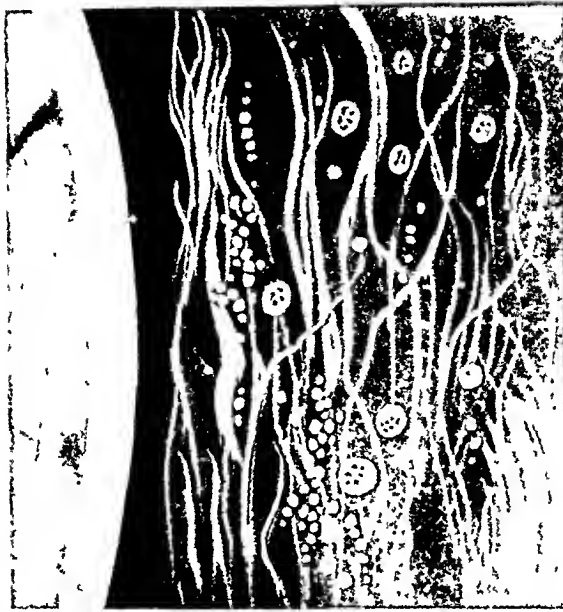
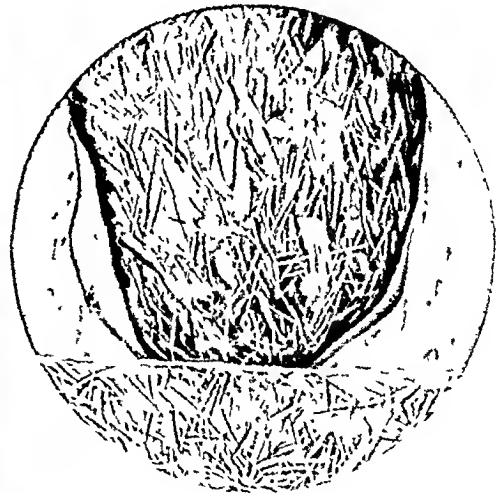


FIG. 503. Asteroid hyalosis. Patient aged 60, vision 0.6. The lens is normal. Note that the filaments are of low luminosity. They are of irregular length and caliber. Numerous white and very luminous opacities (round to oval) arranged in places like clusters. Those out of focus seem larger and fainter owing to circles of diffusion. (After Kobay.)



A



B

FIG. 504. A. Synchysis scintillans. B. Synchysis scintillans in an aphakic eye following severe hemorrhage which occurred five days postoperatively.

gravitate to the bottom of the degenerated vitreous during rest. According to Duke-Elder, they may be likened to "a shower of glittering golden sovereigns or silvery tinsel-like particles." "Ophthalmoscopically . . . they present a beautiful and quite characteristic picture as they flash by with a gleam as of burnished gold." With the biomicroscope the silvery tinsel-like appearance is mostly seen. This picture is reminiscent of the snowstorm effect created in the old-fashioned paper weights (Fig. 504 A, B). In synchysis scintillans the beam also reveals considerable disruption of the vitreous framework with the formation of relatively dark empty spaces.

CHANGES IN INFLAMMATION

Similar to the lens, the normal vitreous is avascular, but, unlike it, it is acellular. Hence the effect of inflammation can only be passive and brought about by the surrounding tissues (retina and uvea). Inflammation in these tissues will induce vitreous changes very early. The term "hyalitis" although apt, is not strictly in accord with the ending "itis" as commonly applied. The suggested term "hyalosis" may be preferable. In the early stages of intraocular inflammation the vitreous is permeated by the outpouring of inflammatory products. Some authors have described an immediate haziness of the vitreous due to a "plasmoid" exudate which increases the tyndall effect. Even at this stage, with sufficiently high magnification and light intensity the tyndall beam will reveal white dots (cells) within the haze. The main differentiation between early inflammatory states and the purely degenerative lesions (myopia, senility) lies in the fact that the former is characterized by an immediate cellular infiltration migrating from the neighboring tissues. In the latter (see above) the changes produced are of an endogenous nature resulting from the degradation of the vitreous colloidal structure. However, as inflammatory processes continue, and bearing in mind the physico-chemical instability of the vitreous, the initial cellular infiltration will be followed by destruction of the vitreous framework, with the production of endogenous opacities. In the end this can surpass that seen in the degenerative states.

A



B

FIG. 505. A. Cells in the vitreous (early sympathetic ophthalmia). B. Keratic precipitates. Flare in the anterior chamber and deposits on the anterior lens capsule and in the vitreous of the same eye as shown in A.

Among the variety of vitreous opacities seen are:

I. Opacities due to cellular infiltration

A. Dust: dotlike in nature which can aggregate into

1. Larger plaques
2. Comet-shaped and filamentary forms
3. Striae — forming linear, striate, or bandlike configurations

B. Pigment particles

- II. Opacities due to endogenous destruction of the vitreous framework
- A. Flocculations — appearing as tufted or nodular condensations
 - B. Fibrillary or microfibrillary degeneration
 - C. Pseudomembranous formations



FIG. 506 Unusual condensations of exudates in the vitreous in the case of tuberculous uveitis.

The early changes in the vitreous brought about by inflammatory reactions consist in the deposition of small white dots (cells) (Fig. 505 A). Each cell is seen as a brilliant dot by virtue of its diffusion or diffraction of light but the morphology is indeterminate, so differentiation between the cells is impossible. With increasing deposition the vitreous framework may be delineated more clearly by attachment of the cells to the fibrillae. The appearance and distribution, similar to the condition in the aqueous, does not permit of a specific etiologic diagnosis, nor does any specific disease evolve a characteristic picture. Early in the course of an inflammation, depending on the severity, only a few scattered dots may be found in the retrolental space. These cells can only be seen biomicroscopically. As the inflammation progresses, the cells tend to ag-

glomerate into larger clumped forms, thus producing plaques — (stellate or comet-shaped opacities or filamentary forms) — (Figs.



FIG. 507. Comet-like condensations of exudates in the vitreous in uveitis.

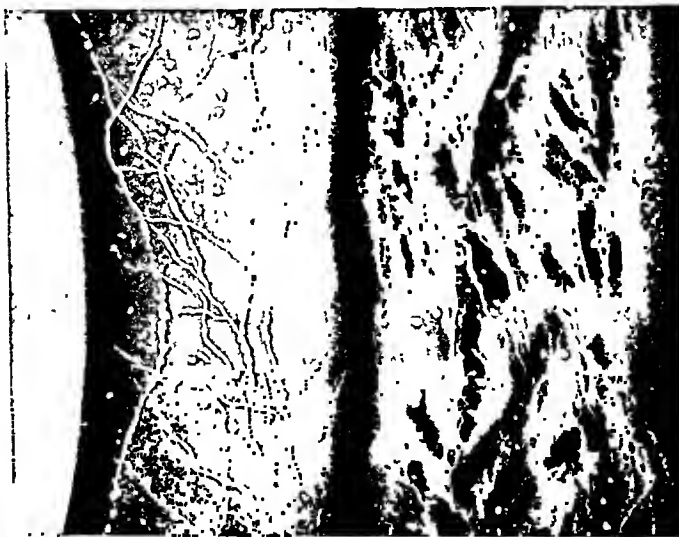


FIG. 508. Deposits attached to the vitreous framework and arranged in linear and striaform configurations.

506-508). A yellowish tinge may be imparted to these denser collections. These latter may coexist with the early discrete dots.

Arrangement into linear, striaform or banded configurations of dot opacities is seen, frequently resembling a galactic "milky way" (Fig. 508). Such a distribution may be, in part, governed by the

PLATE LXXXI

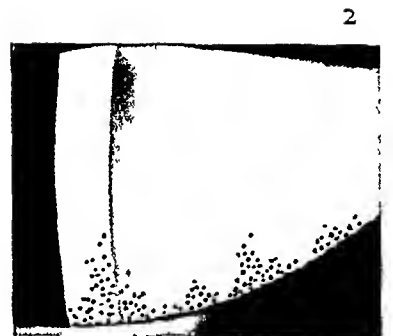
FIG. 1. Postinflammatory pigmentation of the vitreous (after uveitis).

FIG. 2. Same case as shown in Figure 1. Illustrating the pigment deposited on the anterior lens capsule.

FIG. 3. Pigment deposits and vitreous changes in a case of detachment of the retina. Early signs of complicated cataract.

FIG. 4. Pigment deposited in the vitreous (sympathetic ophthalmia).

FIG. 5. Pigmentation of the vitreous in retinitis pigmentosa. Complicated cataract.



structural lamination of the vitreous. These latter are found mainly in the chronic types of disease. Frequently the disrupted vitreous framework is so heavily dusted with a coating of imperceptible particles that an appearance of opaque veils or pseudomembranes results (see Figs. 496 and 497).

It is the author's impression that depending upon the nature and intensity of the disease process uveal pigment granules may be discerned very early in acute fulminating attacks or they may appear also as a late manifestation in the chronic forms (Plate LXXXI, fig. 1). Although the individual dots as such cannot be recognized ophthalmoscopically, when they are numerous and disseminated a reddish hue is imparted to the nervehead while the usually bright-red fundus reflex becomes rosy in tint. This optical phenomenon results from a diffraction of the rays of light causing a shift in the spectrum to the warmer side. However, larger aggregates can be made out ophthalmoscopically as blackish or grayish clumps or strands defined against the reddish fundus background. It is only in synchysis scintillans and asteroid hyalitis that the deposits are sufficiently refractile to gleam yellowish or golden in the former, and whitish in the latter.

It should be noted that the transition of exogenous infiltration to the formation of endogenous opacities (resulting from destruction of the vitreous framework) is not sharply demarcated. Rather the latter formations are indicative of intensity and chronicity of the involvement. The endogenous opacities do not differ in appearance from those seen in myopia and senility (page 1406) except that the concomitant associated cellular infiltrations become more extensive. The condensations and pseudomembranes produced as a consequence of alterations of the vitreous structural framework are more mobile than the earlier punctate precipitates. This results from diminution of the vitreous viscosity. This may be evidenced by the formation of relatively dark spaces or clefts seen between the more reluctant formations. As Duke-Elder⁴¹² describes it "the picture may be complicated; an abundance of minute white specks, spangles and threads are mingled with large agglutinations like tangled skeins of wool,



FIG. 509. Vitreous deposits. Cellular deposits in the vitreous with early disruption of the framework. Iridocyclitis.



FIG. 510. Cellular deposits in the vitreous with more advanced disruption of the framework. Right eye; heterochromia syndrome. The iris is discolored. Numerous keratic precipitates present. Vision 0.9. Posterior cortex of the lens showed a polychromatic luster (irridescence) owing to beginning of cataracta complicata. Anterior parts of the vitreous were dusted over with fine white opacities. Further back a dark space containing vascular remnants is seen. Still deeper are dusty pseudomembranes. (After Koby.)



FIG. 511. Cellular deposits in the vitreous with marked relucency of the disturbed elements of the vitreous framework. Iridocyclitis. Female, aged 20. Probably tuberculous. The posterior capsule of the lens shows a few striaform deposits. The whole of the vitreous structure is accentuated (more relucant) owing to dust adherent to the fibrous type of framework.



FIG. 512. Chronic iridocyclitis. Very extensive infiltration of cells. The vitreous framework is disrupted and owing to a heavy dusting has become opaque, giving the impression of being formed of veils or membranes. Beginning cataracta complicata. Smudged area above the lens opacity is caused by iridescence, the color display exhibiting green, red, and yellow hues.

broad sinuous bands, wavy membranes, and extremely dense floating clouds, the whole oscillating slightly and swaying across the pupil with movements of the eyes" (Figs. 509, 510, 511). Finally, after a

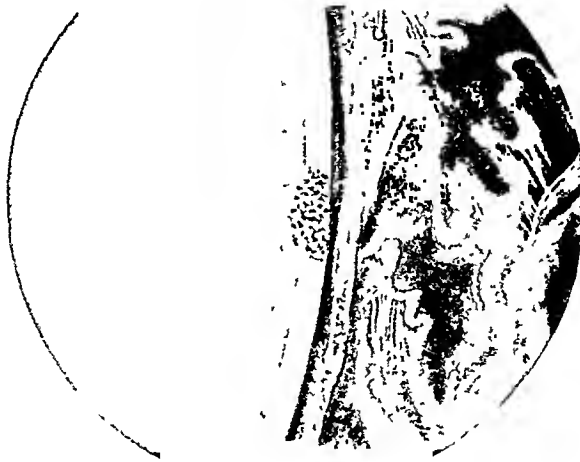


FIG. 513. Pseudomembranes in the vitreous in chronic uveitis. Anterior separation of the vitreous. Deposits on the vitreous surface almost gives the impression of a reduplication of the stripe of the posterior lens capsule. The vitreous has been altered so that its structure appears to be formed of very luminous whitish pseudomembranes in a fanlike design. The increased relucency of these structures results from a dusting by fine deposits.

protracted iridocyclitis and consequent to the formation of dense white membranes with marked cellular infiltrates the opacification of the vitreous becomes so great that these structures may be seen ophthalmoscopically (Fig. 512). In these cases, as reported by Comberg (1924), an anterior separation of the vitreous from the lens has been noted. This may be indicated by the fact that the anterior surface of the displaced vitreous becomes encrusted heavily with fine whitish deposits (Fig. 513). According to Koby, no characteristic vitreous changes are found in glaucoma. Whereas in optic neuritis (papillitis), in sympathetic ophthalmitis, and at the beginning of uveitides, the tiny opacities are not too numerous. They are innumerable in the choroiditides, the chronic uveitides, and the pigmentary retinitides. Generally, they are not seen in the "circinate" or "albuminuric" types of retinitis, the embolizations or thromboses of the retinal vessels nor in the retrobulbar or toxic neuritides.

While an association between the appearance of opacities and the etiology of the underlying condition is not possible, certain special conditions deserve separate consideration.

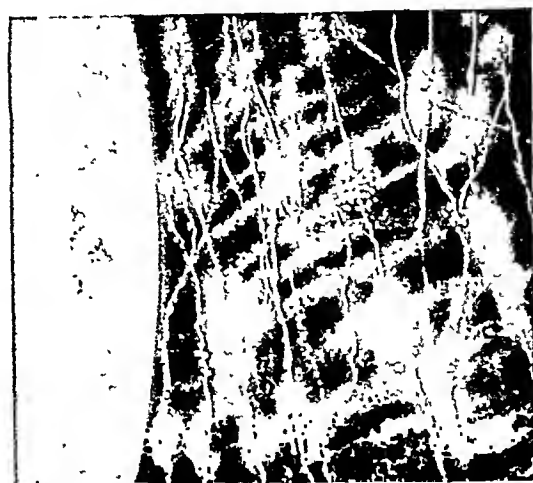


FIG. 514. The vitreous in heterochromic cyclitis. Note the star-shaped opacities in the vitreous and the precipitates in the posterior lens capsule. (After Vogt.)

HETEROCHROMIC CYCLITIS

Since the work of Fuchs (1906) this disease has been recognized as a special type of lesion characterized by an unusual combination of a low intensity process leading to iris color changes, absence of synechia, and the presence of posterior corneal precipitates without visual loss, until late development of complicated cataract. A feature which is found frequently in the vitreous is a disseminated dust-cloud of dotlike or star-form opacities or white to yellowish plaques. As Vogt (1921) significantly pointed out, it may occur without alteration of the vitreous structural framework (Fig. 514). Precipitates occur also on the posterior lens capsule. Unilaterality of involvement is most frequent and the sequelae are complicated cataract or secondary glaucoma in many instances. In a case described by Koby occurring in a 33-year-old male with blue irides, the left eye contained some diffuse yellowish pigment. The anterior vitreous was studded with very fine white opacities, a dark space containing embryonal vascular remains occurred next and the deeper vitreous was regularly stratified with white opacities of variable size. On the posterior lens capsule whitish deposits were seen.

SYMPATHETIC OPHTHALMITIS

The vitreous in sympathetic ophthalmitis was studied biomicroscopically by Vogt (1921). He held that "occurrence in the aqueous,

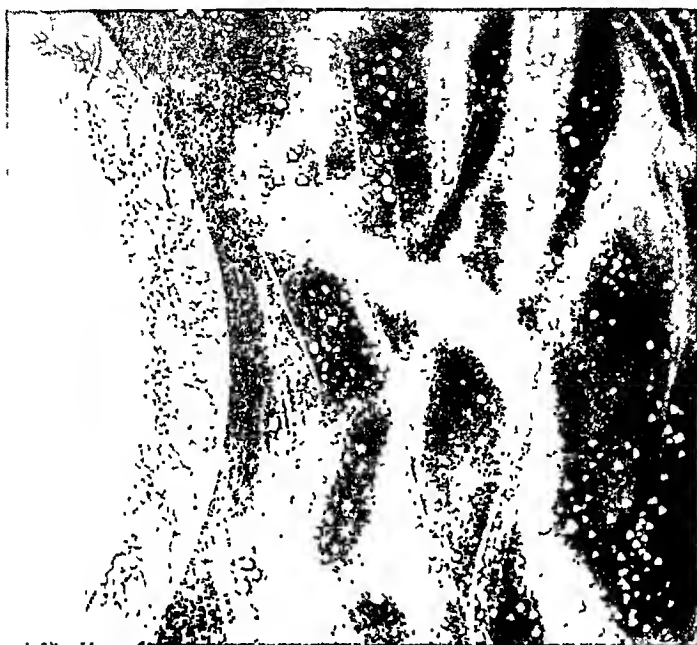


FIG. 515. The vitreous in sympathetic ophthalmitis. A case of a young Negro woman who six weeks previously had had an operation (iridencleisis) for chronic simple glaucoma in the other eye. Note the heavy exudate on the posterior lens capsule.

retrolental space, and the vitreous of free-floating particles (whitish, reddish, or brown) often in large quantity" are invariable early phenomena (Fig. 515). In ephemeral reactions of the sympathizing eye (subsiding in a few weeks) whitish and brown lines and opacities (white dots and pigment) of a transitory nature occur. It has also been noted that the opacities were especially numerous in the lower portions of the vitreous. Minute, repeated accurate biomicroscopic examination of the sympathizing eye is a necessity in all suspected cases. Biomicroscopically, it should be pointed out that there are no clear delineations between sympathetic irritation and sympathetic ophthalmitis *per se*. (See chapter on iris — Sympathetic Ophthalmitis, page 905.)

The insidious onset and the early absence of all signs, except retrolental opacities of a whitish brilliant nature (which when

marked may form a carpeting on the posterior lens capsule) are deceptive, as Molotese (1924)⁵¹⁷ has stressed. Deposits on the posterior lens capsule may be fixed or if they are attached (stringlike) by one end, they may be slightly motile. With aggravation of the sympathetic process this disease presents a picture of marked infiltration and endogenous destruction of the vitreous common to all severe and fulminating intra-ocular inflammations.

OPTIC NEURITIS (PAPILLITIS)

It is only in definite inflammation of the optic papilla that one can expect to find vitreous opacities. They are not seen ordinarily in retrobulbar and toxic affections of the optic nerve. Koby⁵⁰³ has pointed out that in inflammations of the papilla the opacities may emigrate anteriorly via Cloquet's canal where they can remain localized. The clinical findings, in the beginning, may include a "cloud of opacities in the posterior vitreous."⁴⁰⁸ In later stages of optic neuritis fine deposits of pigment upon the vitreous fibers may be discerned. Koeppe has suggested that inspection of the posterior vitreous (by means of a contact glass) may aid in the differentiation from papilledema by finding these deposits present in the earliest stages.

It has been observed occasionally that the occurrence of cryptogenic vitreous opacities as clouds of fine dots appearing suddenly (sometimes quite intensely) may be associated with ophthalmoscopically invisible foci of the uvea.*

RETINAL DETACHMENT

The vitreous in spontaneous retinal detachment as a rule displays extensive alteration both of the exogenous and endogenous types. Considering the incidence of concurrent myopia and retinal detachment it is to be expected that endogenous vitreous changes might

* Duke-Elder⁴¹³ has stated that "such a happening is of common and general occurrence but is seen most typically in women of late middle age. . . . The importance of the syndrome is that it may well form the prelude to a series of similar attacks each involving the disability of further vitreous opacities, or to more serious and widespread uveal inflammation of a recalcitrant and chronic nature unless the etiology is discovered and combatted—in many cases a task of no small difficulty."



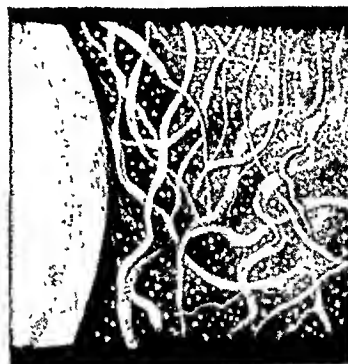
A



B



C



D

FIG. 516. A. The vitreous in detachment of the retina. The detachment followed a dissection of a delicate secondary membrane. B. Vitreous in the detachment of the retina. Disruption and thickening of the framework. Note the clumps and the attachment of vitreous strands to the posterior lens capsule. C. Veil-like opacity in the vitreous following detachment of the retina. Seen soon after retinal separation. The vitreous in the retrolental area is fibrillous and of low luminosity. The deeper layers are more translucent. Below is an area of microfibrillary degeneration. Above there is an ill-defined curved opacity (resembling a dense veil with its convexity pointed downwards) extending posteriorly into the deeper parts of the vitreous. D. The vitreous in detachment of the retina. High-grade myopia. Deposits on the posterior lens capsule. The vitreous shows presence of translucent cylindrical or pointed tubes or cords of irregular caliber, crisscrossing one another, with brownish and whitish deposits. Between them are numerous large round opacities. (After Koby.)

occur. This is demonstrable by condensation of the vitreous framework; thickening of the fibrillae interposed with dark spaces. Progressively such changes lead to the formation of swirling pseudomembranes intermingled with thready fibrillae associated with more delicate microfibrillary degeneration (Fig. 516 A, B, C, D). In addition to these mobile opacities formed by the aforementioned condensations, whitish-brown and reddish clumps or disks (which may or may not be pigmented) are seen. In older cases these may appear as brilliant gilded platelets. Opacities containing or consisting of pigment are characteristic of cases of retinal detachment sometimes appearing in the form of clusters or as isolated dots (Plate LXXXI, fig. 3). Koby has described a special alteration of the vitreous structure in the form of translucent, elongated cylindroids, the ends of which may be pointed, scattered and anastomosed irregularly. An impression is gained that they may be constituted by the curling of a fine membrane. On these cylindroids, one may be able to detect a fine whitish striation or occasional whitish deposits. The complexity of the picture is further added to by the sequelae of acute inflammatory processes (iritis) so common after retinal separation (Fig. 516 D).

RETINITIS PIGMENTOSA (TAPETO-RETINAL DEGENERATION)

As a widespread primary pigmentary retinal degeneration with tapeto-retinal atrophy, this disease presents a characteristic clinical picture, which inexorably progresses to great loss of visual function. It may well be that the degenerative process tends to elaboration of toxic substances into the vitreous. Such toxins may also play a role in the ultimate development of a posterior polar complicated cataract commonly seen in this condition. The degree of vitreous change varies from case to case and usually is a later manifestation. Vogt described a case of a 39-year-old man in which grayish-white ropelike strands occurred in the lower vitreous with discrete white dots deposited on the vitreous framework. Also white cloudy cotton-like agglutinations of the structure were seen as if the latter was

filled with a fine white dust. Pigment in the form of dust or dots may be found in the vitreous or on the posterior lens capsule (Plate LXXXI, fig. 5).

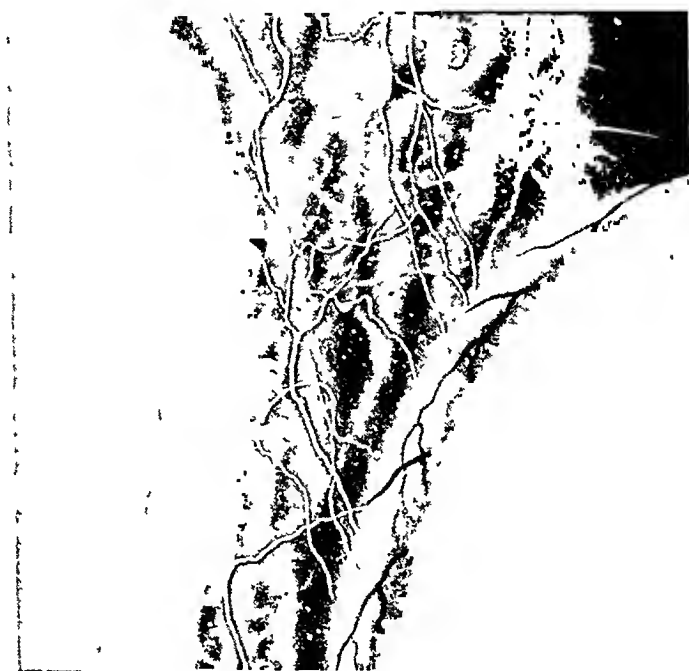


FIG. 517 Melanosarcoma protruding into the vitreous just behind the lens. Note vessels overlying tumor mass. The vitreous framework is disturbed and shows infiltration with white and pigmented cells.

TUMORS AFFECTING THE VITREOUS

GLIOMA RETINAE (RETINOBLASTOMA)

In Vogt's cases, in the first of which the tumor had extended anteriorly within the vitreous almost to the lens, rounded, coarse, white and brown precipitates could be seen on the surface of the tumor and the posterior lens capsule. In the second instance, the vitreous was filled with freely floating dustlike scintillating deposits. Similar ones were seen adherent to the posterior lens capsule. Vogt attributed the colored scintillation to fine cholesterol crystalline deposits. Considered of great diagnostic importance is the finding of detached clumps of tumor cells in the vitreous. Ophthalmoscopically these may appear as large floating opacities.

MELANOSARCOMA

In certain cases of melanosarcoma, particularly those of the ciliary body, the protrusion of the tumor may be so far forward that the dark brownish surface falls within the scope of the beam (Fig. 517). When this occurs, it is often possible to make out certain details of the surface of the mass. Sometimes the malignant tumor in the ciliary body may compress the vitreous against the lens. Occasionally in such cases some brownish points may be seen in the area occupied by the compressed vitreous. It is not possible definitely to state whether these are metastatic tumor cells. In other cases the tumor may actually indent the lens itself.

In a personal observation a delicate, fuzzy, whitish layer was seen to cover the dark brown pigmented tumor mass. This may have been degenerated retinal tissue. Also in many cases the retinal vessels may be seen coursing over the tumor. Disruption of the vitreous frame with varying amounts of cellular infiltration, both white and pigmented, is common (Fig. 517).

TRAUMATIC INJURIES TO THE VITREOUS

From the viewpoint of the biomicroscopist injuries to the eye, whether due to external violence (contusions, or perforations with or without presence of foreign bodies) or operative interference may produce the following changes in the vitreous:

- I. Increase in relucency of the tyndall beam
 - A. Plasmoid vitreous
 - B. Hemorrhage
 1. On posterior lens capsule (retrolental space, Berger)
 2. Intravitreous
 - a. In Clouquet's canal
 - b. Laminated or parallel bands or striae
 - c. Disseminated
 - C. Pigment (uveal)
 - D. White cellular elements
 - E. Lenticular or capsular debris or residua

- II. Disruptions of the vitreous framework
 - A. Traction bands
 - B. External prolapse
 - C. Internal prolapse or herniation
 - D. Kerato-vitreous adhesions or synechiae.
- III. Visible foreign bodies



FIG. 518. Starlike figures in the vitreous following trauma (nonperforating injury). These figures are the result of the deposition of formed elements (white cells) on the fibers of the vitreous framework.

INCREASE IN RELUCENCY OF THE TYNDALL PHENOMENON

One of the most elusive reactions in the vitreous, seen especially after contusion, is an increase in relucency of the focal beam. This is the result of a plasmoid effusion. It may be the only sign following a contusion and is demonstrable only biomicroscopically. However,

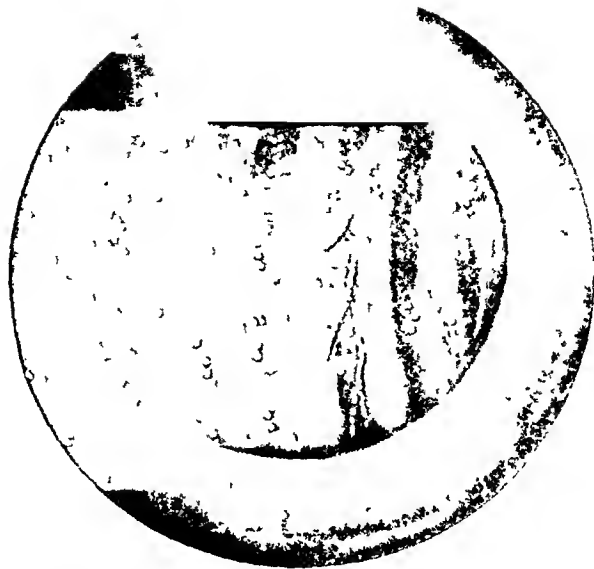


FIG. 519. Increased lucency of the vitreous tyndall. Aphakia; cyclitis. Note flare and cells in the aqueous. The focal beam is very reluctant as it passes through the vitreous owing to plasmoid effusion and cells. The vitreous stratification is preserved. (After Koby.)



FIG. 520. Vitreous hemorrhage following trauma. Note striate-like deposits on the posterior lens capsule. Blood cells are seen disseminated throughout the vitreous. Most of them are adherent to the framework. The color of these cells (not reproduced here) was lemon yellow.

PLATE LXXXII

FIG. 1. Old hemorrhages in the vitreous. Blood cells deposited on the vitreous framework. Posterior complicated cataract.

FIG. 2. Hemorrhage in the vitreous. Fresh blood seen above outlined by the upper border of the hyaloid. Below the older blood cells are yellow in color.

FIG. 3. Pigment deposits in the vitreous and on the posterior lens capsule. Uveitis.

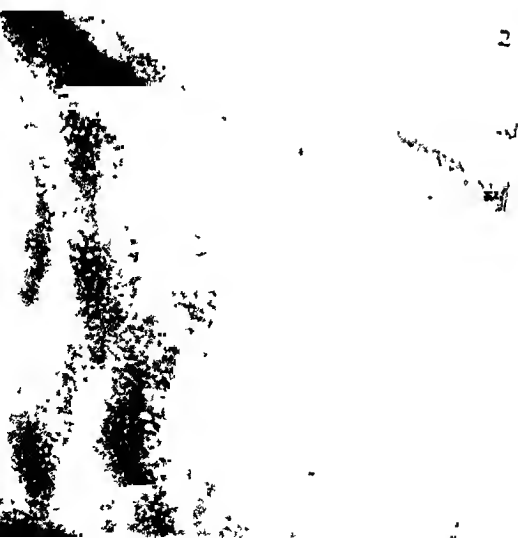
FIG. 4. Hemorrhage in the vitreous observed one year later. Note the lack of color in the cells.

FIG. 5. Hemorrhage in the vitreous. Fresh blood in the vitreous and on the posterior lens capsule. (After Koby.)

FIG. 6. Blood cells outlining prolapsed vitreous into the anterior chamber. Hemorrhage into the anterior chamber occurred four days after an intracapsular cataract extraction.



6



4



3

when it is marked, its presence may be suspected ophthalmoscopically by a careful observer, owing to slight blurring of the fundus reflex and details. Depending on its degree, such a change may be ephemeral; but marked increase of formed elements gives rise to residua which may remain for long periods (Fig. 518). As described previously, increase in the tyndall effect is seen best by observing the passage of the focal beam into the vitreous with the unaided eye (fentoscopy). This increase in the tyndall effect is produced by a combination of formed and unformed elements, probably fibrin coagulants intermixed with whole or fragmented cells, or the increased light dispersion may be caused wholly by liberated pigmented debris (uveal) (Fig. 519). Very often in contusions the development of a plasmoid aqueous (see Vol. I, page 588) may hinder observation of the vitreous because of a screening effect.

Hemorrhage

Hemorrhage into the vitreous is probably the commonest finding of all after injury (Fig. 520). It may be seen adherent to the posterior lens capsule, in the retrolental area or within the definitive vitreous substance (intravitreous). It should be kept in mind that hemorrhage may be the consequence of pathologic processes posteriorly and from the biomicroscopic appearance alone differentiation may not be possible. In this connection Ascher (1920)³¹⁹ pointed out that in cases of trauma the hemorrhages that come from the ciliary body tend to diffuse into the anterior vitreous and especially upon the posterior lens capsule.

On the posterior lens capsule the hemorrhagic deposits are bright red in color with the focal beam, forming sinuous bands, which may be delineated inferiorly as crescentic curves by the ligamentum hyaloidea-capsulare (Weigert). This color is best seen with the unaided eye. Depending on the amount, distribution and duration, the picture of blood on the posterior lens capsule varies. It may appear also as bright-red plaques, or as pointed out on page 1219, distributed as annular rings, perihyaloidian clumps, within the arcuate line, or disseminated (Koby). (See Plate LXXXII, fig. 5.) According to its

duration, the blood depositions may change in character and color owing to laking and disintegration. In such instances the color of the deposits on the posterior lens capsule may change from bronze



FIG. 521. Striate deposits on the posterior lens capsule following trauma (contusion). Young man injured one day previously by a cut wire which struck the center of the cornea. Posterior surface of the lens shows numerous deposits in the form of irregular striae, giving the appearance of moss. The relucency (tyndall) of the vitreous is increased. Note rests of the hyaloid artery (corkscrew-like filament) attached to the posterior lens capsule. (After Koby.)

(similar in appearance to Vossius' ring) to yellow and eventually to a whitish glistening dust. Sometimes overlying the cellular deposit on the posterior lens capsule, white cloudy epicapsular depositions (fibrinous residua of blood) are discerned (Fig. 521). In flat depositions, fixation of the blood elements to the posterior lens capsule occurs (Plate LXXXII, fig. 5). According to its quantity, the deposit bulges posteriorly into the retrolental space like a cap thinner peripherally than centrally, and may appear to move slightly with eye or head movements. Evidences of hemorrhage on the posterior lens capsule and within the retrolental space may remain for extensive periods.

Intravitreal hemorrhage. Hemorrhages within the vitreous proper present varying appearances according to the individual circumstances concerned. By the time the case is presented for inspection or biomicroscopic examination is feasible, hemorrhage into the vitreous may be an outstanding feature. The blood cells may be

diffused and disseminated upon the vitreous framework or it will be found regionalized, occupying certain areas, appearing as irregular localized collections (Fig. 520). In other cases, the deposition may be arranged as tensed bands separated from one another by dark intervals (lamination). After perforations, parallel bands of this nature, consisting partly of blood cells and stretched vitreous framework, may be seen extending rectilinearly toward the wound and also may be marked on the opposite side. In a case noted by the author an adherent band covered with blood cells stretched from the lower part of the posterior lens capsule backward into the disorganized vitreous. The retrolental space was obliterated.

Intravitreal collections apparently confined to or outlining Cloquet's canal (or distending or sacculating it) are seen often. Histologically there is confirmation of this possibility not only with hemorrhage but also with inflammatory products (Fuchs; Samuels). Koby cites Hoffman (1926)⁵⁰² who reported the case of a lad of 8 years; two months following contusion, the globe revealed a dark tunnel outlined by reddish points (the anterior extremity of Cloquet's canal) immediately behind the posterior pole of the lens. Behind this the accumulation was so great as to prevent deep penetration of the beam. In pathologic instances, e.g., juvenile retinal and vitreous hemorrhage (Eales' disease), Vogt described several cases in which similar collections of blood were found in the anterior vitreous behind the lens but separated from it. It may be that Cloquet's canal provides a conduit by means of which passage of the blood anteriorly is facilitated.

Vogt reported an instance following perforating injury in which retinal hemorrhages were seen around the papilla. In the vitreous most of the blood was concentrated into linear strands in the anterior portion. These strands moved very slightly. After resorption of the blood elements the coarse strands became finer and seemed to converge to the place of perforation and to the opposite side of the base of the vitreous. The remainder of the vitreous was dusted with glistening erythrocytes. After a year, the strands were still present.

Evidently such strand formation is a characteristic sign in perforating injuries and hence is of diagnostic importance even years after the injury.



FIG. 522. Pathologic vitreous changes following a nonperforating injury. A heavy cellular infiltration is seen throughout. Striate deposits on the posterior lens capsule. Behind the lens there are numerous crossing filaments. Deeper to this area there is a zone of pseudomembranes dusted with opacities. Still deeper there appears to be a traction band.

Pigment

Uveal pigment depositions are seen commonly following traumatism or surgical interference. This is especially true in herniation (see below). The color of these deposits is reddish-brown. As already indicated (page 823) one must exercise caution in basing a differential diagnosis of vitreous conditions on their color alone. Pigment sometimes may agglomerate into larger collections or, being precipitated on preexisting vitreous opacities, may cause them to assume a brownish appearance. Factors influencing the judgment of color are intensity of illumination, magnification employed, screening or masking effect by the intervening tissues (e.g., senile yellowing of the deeper parts of the lens, etc.). Masses of unlaked erythrocytes will appear bright red at first. However, thin blood films similar to a microscope slide smear appear brownish to a light yellow. And even cells that appear lemon-yellow become whitish in arc illumination. Eventually, laking of the blood pigment causes loss of

the yellow color, even using the ordinary nitra beam so that differentiation of red cell derivatives from inflammatory cells is not always possible.

In the main it may be said that fresh red blood cells in aggregate will appear reddish. When disseminated or laked they will appear lemon-yellow in coloration and eventually white. Uveal pigment, on the other hand, appears reddish-brown and tends to persist so.

White Cell Elements

In exudative phenomena white cells may be seen early in the plasmoid vitreous. They lie suspended upon the vitreous framework and differ in no way from those in inflammatory conditions. Deposits in the form of white dots or striae resembling moss may also be seen on the posterior lens capsule (Figs. 521 and 522).

Lenticular Debris

Following perforating injury or surgical procedures in which the lens capsule is ruptured and the lens material is macerated, fragments of cortical lenticular substance and even capsular remnants may be incorporated within or lie upon the vitreous mass. In these cases prolapse into the anterior chamber is common.

They appear as ill-defined intensely white granular fragments or portions of longer lens fibers incorporated within the surrounding disrupted vitreous.

DISRUPTIONS OF THE VITREOUS FRAMEWORK

In addition to the above-mentioned changes, trauma may affect the structure of the vitreous by causing the formation of local alterations within the framework itself, e.g., in perforating injuries it may be possible to discern a track indicating the course of a penetrating foreign body by the localized vitreous disturbance induced or by the outlining resulting from the deposition of cells or of foreign body detritus. In this respect Koby cites the work of Ogawa (1906)⁵⁵⁵ who showed in experimental animals that after introduction of a cataract knife into the vitreous, the fibrillae become con-

densed about the channel of penetration. This channel contained only a clear liquid with several migrating cells, which subsequently disappeared. From this it would appear that loosened fibrillae may retract somewhat and in such cases may favor the migration of a foreign body. Non-perforating contusion also may cause great disruption of the vitreous framework. "Liquefaction" with the formation of large dark lacunal and freely motile fibrils and cloudlike membrane may occur. Bands apparently under traction may also appear. (Fig. 522). However such bands are more common following perforating injuries or operations (needlings) or after extrusion of vitreous from the eye (cataract operations).

Herniation of Vitreous. In this discussion it is not necessary to dwell upon these instances of severe perforating injuries attended by gross extrusions of vitreous from the eye. Our concern is primarily with the intraocular herniations since their frequency and evaluation have been recognized only since the advent of the biomicroscope. The importance of zonular rupture even in minimal injuries was stressed by Frenkel in his conception of the so-called "anterior chamber syndrome" (page 913). In these cases even where subluxation may not be frankly apparent herniation of the vitreous anteriorly may occur. In the more evident lenticular displacements, prolapse of the vitreous into the anterior chamber is very common. In injuries in which iridodialysis results, herniation through the artificial pupillary orifice may be noted as a consequence of zonular rupture.

It is a daily experience with surgeons that herniation of the vitreous is found biomicroscopically after intracapsular cataract extraction procedures and especially following discissions or extractions of secondary cataracts (Fig. 523 A, B). In the latter it is extremely rare not to find some evidence of prolapse.

Ordinarily, on displacement of the lens the vitreous tends to herniate anteriorly in such manner that the anterior surface of the bulge is more or less regular. The biomicroscope will reveal only a minimal indication of the interface between the vitreous and aqueous humors because it reflects so little light. However, by imparting a slow oscillatory movement to the focal beam the tenuous surface of



FIG. 523. A. Herniation of the vitreous into the anterior chamber following discission of a delicate secondary membrane. Surface of the bulging vitreous is smooth and well demarcated. Near the pupillary border the faint remains of the secondary membrane are seen. B. Herniation of fluid vitreous into the anterior chamber following an intracapsular cataract extraction. Smooth surface of prolapsed vitreous is outlined by cells.

the vitreous can be always recognized. Frequently in such cases it will appear as if a hemispherical bulging of vitreous hangs over the pupillary border. Because of the bulging vitreous, the border itself

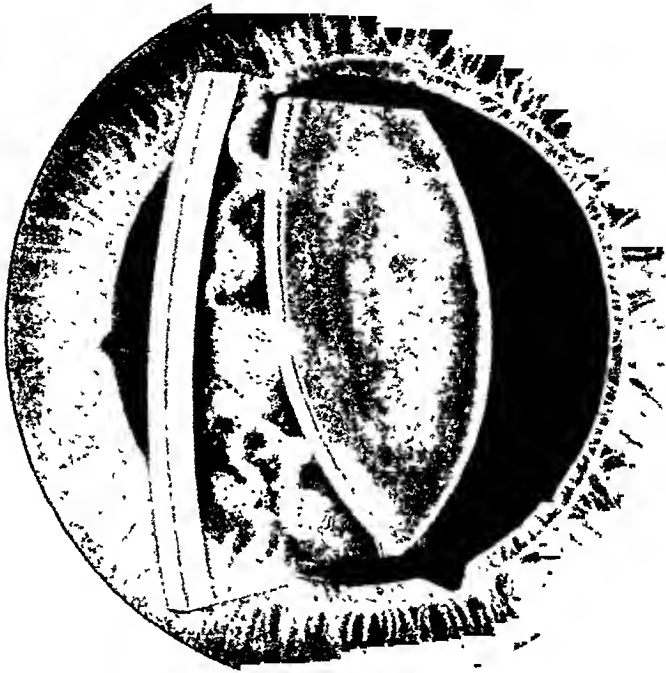


FIG. 524. Herniation of fluid vitreous into the anterior chamber following trauma. Lens is displaced upwards and backwards. Note tear of iris border below.

may be displaced somewhat anteriorly from the lens so that a noticeable separation exists at this point between the iris margin and the lens.

It often happens that following cataract extraction, adherence of vitreous to the iris and to the posterior corneal surface may by organization induce peripheral anterior synechiae and consequent secondary glaucoma.

The bulging vitreous may vary in size from a slight bead or be so extensive as to reach the posterior corneal surface. In some cases (fluid vitreous) the prolapse may be irregular and ill-defined so that no regular surface presents (Fig. 524).

The appearance of the vitreous within the herniating portion will vary from case to case and is composed of irregularly coarsing trabeculae and fibrillae, frequently containing pigment. In other in-

stances streaks of blood cells may be seen especially in the dependent portions. Depending on the consistency, dark or less reflective spaces may be seen within the mass. It has been noted by several writers that the contained blood within the vitreous has little tendency to resorb. Very commonly the lightly reflecting surface of the prolapsed vitreous is defined and made more visible by the deposition of pigment granules on the surface and this frequently makes its presence more easily recognizable. According to Koby, change in the size of the prolapse may occur especially when it is small. He has noted that diminution of size over a period of several months occurs with the smaller hernias. He suggests that the massaging action of the iris on the prolapsed mass is capable of leading to partial reduction in size.

It is interesting to speculate at this point on the nature of the anterior limiting layer of the vitreous. Vogt in his discussion considers that normally the anterior and most highly reflecting band (the plicated membrane) seen behind the retrolental space might be considered as the anterior limiting layer of the vitreous. Since the retrolental space in front of this contains the hyaloid vitreous it would follow in this sense that this part is extraneous to the vitreous proper. If this is the case, does the hyaloid vitreous escape after every total lenticular extraction? It seems to me that the most anterior or plicated membrane does not represent the anterior border layer of the vitreous; that it is the hyaloid membrane itself which probably is in contact with the posterior lens surface, and that only a potential postlenticular capillary space exists. In this case, because of the high degree of reflection of the posterior lens capsule, the anterior limiting layer of the vitreous is not apparent biomicroscopically. Perhaps the only time it is visible is after intracapsular operations and herniations.* As to the tenuous surface of demarcation between the aqueous and herniating vitreous, Duke-Elder¹¹⁴ states: "The apparently distinct membrane is probably a mechanical effect of the tension and

* Following intracapsular cataract extraction, where herniation of the vitreous is commonly seen, rupture of the "hyaloid" and spilling of vitreous substance into the anterior chamber may occur spontaneously months postoperatively. This phenomenon may be attended by slight ciliary injection. Improvement in visual acuity has been noted following such an episode.

stress created by the hernia on the micellar structure of the gel, causing a rearrangement, a reorientation and a condensation of its fibrillar elements."

Would this process be instantaneous? Following a traumatic herniation into the anterior chamber, I have seen within two hours a typically outlined vitreous bulge with a surface such as that described above. Similarly, on the day following a discission of a secondary membrane (where the hyaloid membrane was certainly incised) a smooth bulging and sharply delimited herniation of interior into the anterior chamber was observed. And how shall we explain the cases of vitreous prolapse following discission, trauma or intracapsular cataract extraction in which an irregular extrusion of vitreous occurs and in which no smooth or rounded delimiting surface is seen? Such differences in the appearance of herniated vitreous probably depend on its physical aspects; thus representing in one case a more or less turgid gel-like consistency and in the other a higher degree of fluidity. Practically, this has been well known for a long time to surgeons when witnessing vitreous escape during operative procedures. It will take considerably more experimental investigation before one can give any definite opinion concerning this problem. Perhaps if some specific method of intravital or histologic selective staining could be devised, light might be shed on these questions.

Larger hernias seem to persist indefinitely. The prolapsed vitreous trembles somewhat upon movement of the globe. In several of Koby's cases he noticed a sagittal oscillation accompanying pupillary movements. St. Martin describes also a pulsation synchronous with the arterial pulse.

Several authors, St. Martin, Poyales and Moreno (1934),⁵⁶⁴ in their studies of prolapse after total extraction of the lens, found a very high incidence of anterior prolapse of the vitreous. According to these writers it will be seen at times that the "hyaloid" is torn as indicated by irregular borders. In other cases a large hemispherical bulging of the vitreous will be observed; its surface may or may not be marked by deposits. Herniation or prolapse of the vitreous

into the anterior chamber during or following extraction of the lens may be a factor in the causation of delayed restoration of the anterior chamber, distortion of the pupil, anterior peripheral synechiae and

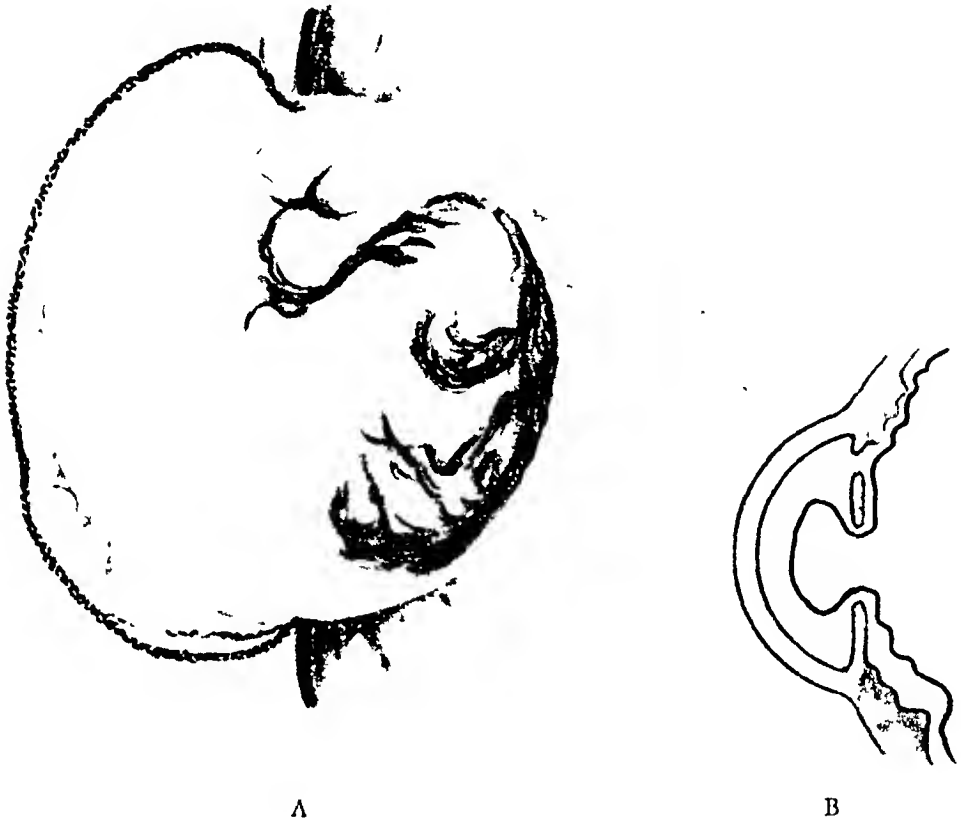


FIG. 525. A. Herniation of formed vitreous into the anterior chamber following an intracapsular cataract extraction. Observe the sharp outline of the vitreous surface to the left. Deeper veils of disturbed and relucient vitreous are seen. (After St. Martin.) B. Diagram of A.

consequent secondary glaucoma. This may result from the organization of vitreous bands (Figs. 525 A, B; 526 A, B).

When loss of vitreous occurs during an intracapsular extraction bands of vitreous may be found drawn up to the corneal wounds. Similar bands are often seen following discissions of secondary membranes. After the extrusion of any considerable amount of vitreous from the eyeball, especially if it is "formed" or nonfluid, opacification of the retained vitreous will occur. This opacification results from cellular infiltration and condensation of the framework.

Prolapse or loss of vitreous following extracapsular operations may result in a mixture of soft lens matter and vitreous. In such cases the intermingling of vitreous material will hinder the absorption of lens cortical substance and may contribute to untoward sequelae.



FIG. 526. A. Herniation of fluid vitreous into the anterior chamber following intracapsular cataract extraction. Note the irregular masses of vitreous protruding into the anterior chamber. No demarking surfaces are seen. B. Diagram of A. (After St. Martin.)

After extracapsular cataract extraction and in the absence of a dense secondary membrane, the posterior capsule can be differentiated easily from the vitreous behind it. With the narrow beam the posterior capsule always appears as a definitive line.

As mentioned above, prolapse of the vitreous always occurs following discission of a secondary cataract. The surface of such a prolapse may or may not be outlined sharply. Sometimes the surface is composed of irregular fluffs and strands and in other instances it is more sharply delineated by the presence of pigment or small whitish

precipitates. Dejean, (1925),³⁹⁶ St. Martin (1930-1935), and Koby (1939)⁵⁰³ believe that the delineation of the prolapsed surface (which some have held is due to a kind of condensation) is due rather to a pathologic change dependent on precipitation.

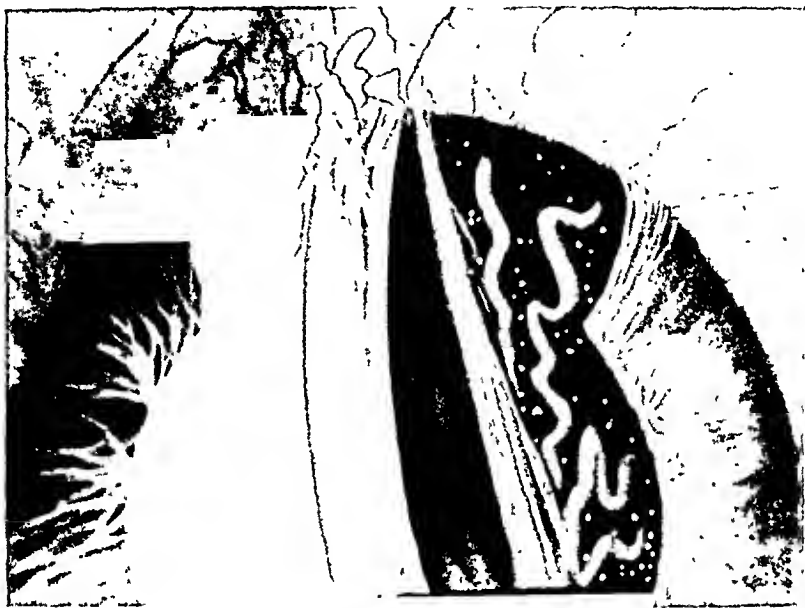


FIG. 527. Keratovitreal synchia. Vitreal and some capsular remains drawn up into the corneal wound following discission of a secondary cataract. Observe the disturbed vitreal (pseudomembranes and cellular deposits).

Not uncommonly after discissions, a tract of vitreal may be drawn up to the corneal wound to which it may become adherent forming a kerato-vitreal synchia. That this occurs very frequently after discission has been brought out by biomicroscopic examination. At the point of corneal adhesion, disturbances of Descemet's membrane may occur (opacifications, folds, and tears). It has also been shown by Methy (1924) that such kerato-vitreal synchia may be established following electromagnetic extraction of a foreign body. Apparently in discission the vitreal is drawn up by the knife-blade as it is withdrawn. Adhesions of this nature are permanent (Fig. 527).

FOREIGN BODIES IN THE VITREOUS

All manner of foreign bodies may gain ingress to the vitreal as a result of a perforating injury. The tract may be evidenced by

corneal or scleral scars. (See Vol. I, page 547 and Vol. II, page 1235.) However, in the case of the scleral perforation it frequently happens that after the absorption of initial conjunctival hemorrhage no evidence of its passage may be found. In the instance of a sterile foreign body its presence may be clearly seen ophthalmoscopically because of the lack of inflammatory reaction provided extensive hemorrhage is not also present. Those lodged in the anterior parts of the vitreous may be seen with the biomicroscope.* To facilitate this the pupil should be dilated maximally.

Foreign bodies distant from the ciliary body or retina are less liable to cause foreign-body reactions than those situated close to those two tissues. Depending on the nature of the substance, the foreign body may be well tolerated in the vitreous for long periods of time. This is especially true of many metallic substances excepting the ferrous and cuprous compounds or their alloys, which rapidly dissolve leading to siderotic and chalcotic changes. Since a larger percentage of the intravitreal foreign bodies are of a ferrous nature the importance of early removal is apparent. The "noble" metals are best tolerated in the order of platinum, gold and silver; the less oxidizable substances such as chromium and aluminum or the silicates are also well tolerated. Since modern alloys vary considerably in their composition no general rules concerning reactions in the vitreous can be formulated. For example, in a personal observation, a piece of brass has been seen in the vitreous relatively unchanged and with no ocular damage or effect upon vision over a span of years. With the ophthalmoscope it appears as a freely mobile golden glittering speck.

As a consequence of war wounds, especially due to mines, the incidence of intravitreal nonmagnetic fragments has been increased. Fragments of greenish plastic from German mines, porcelain, clay or glass from both German and Japanese mines, wooden splinters and gunpowder particles have all been seen at one time or another.

It has been reported that globes containing glass fragments or lead

* Those lying deeper may be seen biomicroscopically by means of the contact lens (mirror or prism) provided they are not located too far peripherally; in this case they may be seen ophthalmoscopically.

granules, after remaining quiescent for a long period of years, may suddenly flare up into activity. In siderosis, Vogt has described whitish opacities of a size varying up to 0.05 mm. Some of the larger ones were pigmented. The remainder of the framework also contained fine brown to reddish opacities.

Of all the common metals, copper is probably the most toxic. In association with chalcosis lentis the biomicroscopic examination of the vitreous may disclose similar (greenish) discoloration of the disrupted framework. Where lead particles have penetrated the vitreous the shiny nature of the globules may be seen for very long periods. Likewise aluminum particles exhibit a shiny appearance.

As a consequence of penetrating wounds, it is obvious that air may be entrapped in the form of single or clusters of bubbles which rapidly absorb. With the ophthalmoscope the appearance of the reflex from them will vary depending upon the direction of light.

It should be recalled that the disruptive influence of foreign bodies causes formation of tracts often outlined by blood cells.

INTRA-OCULAR PARASITES

Parasites invade the eye rarely. Nonetheless consideration must be given the possibility whenever an unusual vitreous cyst or mass is found. Of all parasites, worms and fly larvae are reported as being most frequently found in the eye. The former or nematelmintes (round worm) is represented by filaria (thread worms), and platyhelminthes (cysticercus and echinococcus).

Filarial infestation of the eyes is exceedingly rare insofar as filaria loa, Bancrofti, and medinensis are concerned, but the onchocerca volvulus (microfilaria) invades the eye in most cases and constitutes an important cause of blindness in Africa and the American continents where the organism is endemic.

Infestation by adult forms of filaria have been recorded at times as involving the anterior chamber, lens, vitreous and subretinal space. The motile filaria may be discerned actually lashing about in the aqueous, causing subjective symptoms by the entoptic phenomenon resulting as it whips across the pupil. An inflammatory

reaction, iridocyclitis with cloudy aqueous and vitreous opacities are associated with most cases. Worms first seen in the anterior chamber may subsequently migrate to the vitreous and become lost or die and become calcified. In other instances marked inflammation incites secondary glaucoma and loss of the eye following such migration.

Appearance of filaria in the vitreous must be differentiated from filamentary opacities or hyaloid vascular remnants. Reported appearances are vague. The authentic cases include an instance in which a filaria travelled from the anterior chamber to the vitreous, another in which the organism migrated from a subretinal site to the vitreous chamber and thence to the anterior chamber whence it was extracted via a keratome incision. Kuhnt (1892)⁵¹⁷ reported a case in a 31-year-old man. A pinhead white swelling, paramacular, increased in size in two months and a worm protruded into the vitreous five months later, movements were discerned in the swelling and following a scleral incision the parasite was removed. It was proved to be a nematode. Barrada (1934) reported a case in a 15-year-old Egyptian.³⁵⁴ A red circular hole was seen near the macular encompassed by a raised darkish zone radiating from which were innumerable reflexes of folds of the internal limiting membrane. The vitreous was clear. The patient saw a long black string waving about. Later the filaria disappeared leaving the central hole and opaque film over the surrounding area. Leber (1914) attributed retinal hemorrhages and exudative retinitis to blockage of vessels by microfilaria of *F. bancrofti*.

In onchocerciasis the invasion of the vitreous chamber by microfilaria has been proved to occur. Inflammatory sequelae are the rule. All parts of the eye may be affected concomitantly or consecutively. Entoptic phenomena due to the lashing motile microfilaria may be noticed subjectively.

Ridley (1945)⁵⁷⁷ in an account of an investigation in the Gold Coast among Negroes living in an endemic area found extensive ocular microfilarial invasion throughout the eye. Although the vitreous was not involved alone in his series of cases he notes that in this disease "In the absence of iridocyclitis clarity of the vitreous

seems to be the rule even with marked choroidoretinal lesions though a few opacities such as one of very general occurrence were seen." Silva (1925)¹⁰⁰ saw microfilaria in the vitreous using a plane mirror and Gullstrand ophthalmoscope. Torres Estrada (1942)¹²¹ records seeing worms swimming freely in the vitreous using a slit lamp with a Koeppe mirror and also by means of an electric ophthalmoscope with a ± 20 or ± 40 lens. This writer asserts that microfilaria are commoner in the vitreous than in the aqueous. Cases of vitreous degeneration of the asteroid hyalitis type have been seen, but while onchocerciasis of the eye is a highly prevalent endemic cause of blindness it is not felt that the vitreous is the most likely site of infestation with the microfilaria. It is to be noted that in some cases in which filaria were demonstrable in the anterior chamber, the anterior segment was normal. This has caused some to believe that the entrance of the worms is via the posterior ciliary vessels.

However, today there is little doubt but that the microfilaria gain entrance into the eye by invading and penetrating the cornea. Reaction in the cornea and within the anterior segment of the eye probably depends on the number of microfilaria present and upon the degree of toxicity excited.

The flat worms (Platyhelminthes) are represented by invasive forms of *Taenia solium*, the *cysticercus cellulosae* of which are carried to the eye hematogenously. The commonest site is subretinal. The life of the organism is two to four years and although initially minute, later accretion in size may lead to destruction of the eye. Duke-Elder¹¹⁰ describes the appearance as a cystic swelling, globular and translucent with its wall studded with fine calcareous granules. Leech-like or heaving movements have been noted. The occurrence in the vitreous is common. Extensive retinal separation may result, possibly associated with migration of the parasite within the fundus. Although originating elsewhere, the organism may migrate to the vitreous chamber, its passage through the retina being attended by a violent reaction with marked formation of vitreous opacities. Arrest during the passage may occur and the head become fixed suprachoroidally. If initially in the retina, it escapes readily into the

vitreous and is easily seen. While in the vitreous it may be well tolerated but an eventual uveitis ensues.

Diagnostic confusion of a parasitic cyst with embryonal rests in

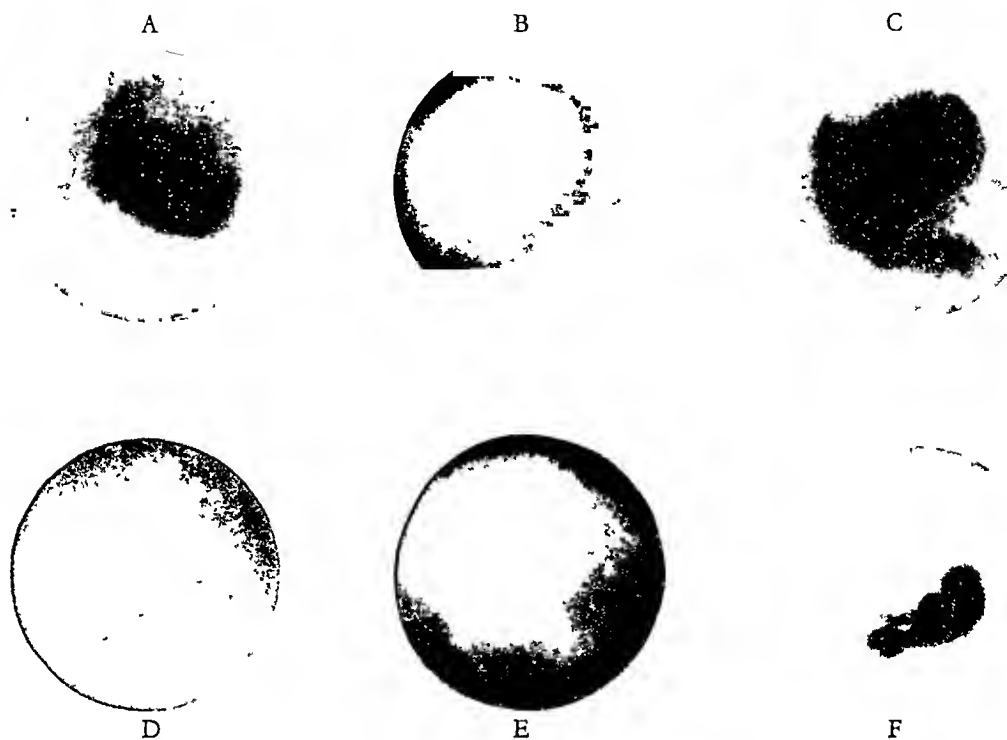


FIG. 528. Free cysticercus in vitreous body. Two of the four suckers may be seen in the lower right picture. (Courtesy of Dr. J. Lech, Jr.)

the vitreous, especially cystic dilation of the hyaloid artery, or with a site of retinitis proliferans (Coats) or with a neoplasm (such as glioma), should be kept in mind. If a cysticercus is suspected, effort should be made to visualize the head (ophthalmoscopically and/or biomicroscopically) or the neck of the organism. The undulatory or peristaltic movements are characteristic but often absent (Fig. 528).

While in the vitreous the preretinal site is commonest; the cysticercus may be more central or it may come closer to the lens. Exceptionally, forward migration to the anterior chamber has been noted. Diagnosis is facilitated by noting the globular translucent swelling formed by the head, intracystic heaving movements and undulations. Stimulation of these movements by temporal galvanization or faradization is suggested. A migratory subretinal cyst is always suggestive of cysticercus. Hooked scalices may be visible. The

biomicroscopic study of a suspected cysticercus is important in any consideration of its removal transclerally to prevent further destruction of the eye. Cosmettatos and Anargyros (1925) recommend subconjunctival injection of a hypertonic saline solution 6 to 10 per cent to clear the vitreous and also to provoke an eosinophilia.

Upon invading the eye *Taenia echinococcus* larvae give rise to a hydatid cyst. Intra-ocular occurrence is rare, rarer than cysticercus. Authenticated instances are uncommon. Gescheidt (1833)¹⁰⁶ described a retrolenticular yellowish opacity. Griffiths (1897) reported a glistening white opaque nonvascular opacity in contact with a clear lens. Werner (1903)¹⁰⁷ found a grayish white mottled opacity behind the lens. A hydatid cyst was found on section. Scholtz¹⁰⁸ reported an interesting appearance in the vitreous of a mass of some 20 small clustered globular vesicles which eventually become obscured by progressive vitreous opacification.

Burrowing within the eye by the larvae of various flies (ophthalmomyiasis interna) has been noted. The incidence among young children is greater than among adults, probably because of the softness of the scleral tunic. Migration to the vitreous evokes considerable reaction, but while in the vitreous less disturbance ensues. DeBoe (1933) reported subsequent larval inactivity, and Anderson (1934) noted shrivelling of the larva without ocular damage. If the larva is seen, diagnosis is evident but if not, it is nigh impossible. Early and frequent biomicroscopic examination of suspected instances is recommended.

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